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Non-paraganglioma tumors of the jugular foramen – Growth patterns, radiological presentation, differential diagnosis

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ABSTRACT

Objective: Most common tumors of the jugular foramen are paragangliomas. However, other lesions, also malignant, may involve the jugular foramen and mimic radiographic presentation of paragangliomas. Therefore, a correct preoperative diagnosis is crucial for best treatment planning.

This study analyzes imaging characteristics of non-paraganglioma neoplasms involving the jugular foramen, with attention given to features helpful in differential diagnosis.

Study design: A retrospective chart search.

Setting: Teritary referral university centre.

Subjects and methods: During the years 1997–2010, 11 cases of jugular foramen tumors other than paragangliomas, with available imaging studies, were identified. Histopathology revealed: 3 schwannomas, 1 malignant schwannoma, 2 meningiomas, 1 hemangiopericytoma, 1 ependymoma, 1 endolymphatic sac carcinoma (ELST) and 2 nasopharyngeal carcinoma metastases. CT, MRI and angiography were assessed to determine tumor growth directions, bone involvement, tumor morphology and vascular composition.

Results: Schwannomas were characterized by parapharyngeal space involvement, jugular foramen expansion, preservation of cortical margins, irregular contrast enhancement. Meningiomas presented diffuse bone infiltration, sclerotic changes, erosion of the cortical bone. Ependymoma showed diffuse skull base infiltration, permeative erosion, heterogeneity, abundant vascularization. Hemangiopericytoma radiologically imitated paraganglioma. ELST showed permeative/geographic bony destruction, heterogeneity, intratumoral bony fragments. Metastases were lytic, solid lesions characterized by circumferential growth, internal carotid artery encasement and stenosis.

Conclusions: A combination of certain radiological features including tumor epicenter, growth vectors, skull base infiltration, bony changes and tumor morphology help

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establish correct preoperative diagnosis and differentiate less common jugular foramen tumors, from most common paragangliomas. Hemangiopericytoma may radiologically mimic paraganglioma.

1. Introduction

Jugular foramen is a complex region of the skull base containing important vascular and neural structures. The most common tumors of the jugular foramen are paragangliomas, arising from the chemoreceptor tissue of the paraganglia. These lesions constitute 60–80% of primary jugular foramen tumors and are histopathologically benign [1,2]. However, other lesions, sometimes of malignant course, may involve the jugular foramen primarily or secondarily and mimic radiographic presentation of most common tumors. Therefore, a correct preoperative diagnosis is crucial for best surgical planning and evaluation of postoperative morbidity and mortality. Radiological differential diagnosis is based on evaluation of tumor features demonstrated by computed tomography (CT), magnetic resonance imaging (MRI) and angiography.

This study analyzed imaging characteristics of non-paraganglioma neoplasms involving the jugular foramen. These findings were compared with radiological appearance of paragangliomas, which constitute a majority of jugular foramen tumors, with special attention given to characteristic features, that might be helpful in differential diagnosis. Usefulness of distinct imaging methods for depicting important differentiating features were discussed.

2. Materials and methods

A retrospective search of the files of the neuroradiology department and surgical database of our tertiary referral university center was performed for the years 1997–2010 and medical records of 51 cases of jugular foramen tumors with available imaging studies were obtained. Clinical and histopathological diagnosis was confirmed and 11 cases of jugular foramen tumors other then paragangliomas were identified.

Histopathologic diagnosis was obtained from surgery in 10 patients and revealed: 3 schwannomas, 1 malignant schwannoma, 2 meningiomas, and single cases of hemangiopericytoma, ependymoma and endolymphatic sac carcinoma. A pathology report was not available in 2 patients with skull base metastases in the region of the jugular foramen. They were included in the study, because metastatic disease was suspected on the basis of radiological findings and clinical evidence – the lesions occurred 3 months and 4 months after radiation therapy for the squamous cell carcinoma of the nasopharynx.

All patients underwent CT and 9 patients underwent MRI. Eight patients underwent digital subtraction angiography (DSA), with a standard technique including selective catheterization of both internal and external carotid arteries, as well as the vertebral artery on the side of the tumor.

Also 10 cases of jugular foramen paraganglioma randomly chosen from the radiology department files of the same time period were included in the study to facilitate highlighting radiological features useful in differential diagnosis. The study was approved by the institutional ethical committee.

All images were reviewed to determine tumor location and growth directions. CT scans were analyzed for types of bone involvement, such as pressure expansion, permeative growth, destructive or sclerotic changes. Special attention was given to bone margins of the jugular foramen. The presence of hyperostosis and calcifications was noted. On MRI tumor morphology was analyzed, including signal intensity, degree of contrast-enhancement, the presence of flow-void areas. On angiographic images the vascular composition of lesions was evaluated, with specific attention given to the presence of vascular blush.

3. Results

Clinical details and summary of radiologic features of jugular foramen tumors are listed in Table 1.

All *schwannomas* occupied the jugular foramen and the parapharyngeal space, with anteromedial displacement of the extracranial portion of the ICA. Involvement of the posterior fossa was observed in 1 patient. On CT scans tumors were characterized by enlargement and sharp contours of the jugular foramen (Fig. 1). Both *meningiomas* were primarily centered in the jugular foramen, extended to the CPA and encased the extracranial portion of the ICA. One ''dumbbell-shaped'' tumor had large extracranial component and abundant intra- and extracranial calcifications. On CT erosion of jugular foramen cortical margins, without its significant widening was a constant finding (Fig. 2). On MRI the intracranial component was a dural-based enhancing mass in one case and in another case had ''en plaque'' appearance (Fig. 3). *Ependymoma* occupied the jugular foramen, CPA and extended to the middle ear cavity, external auditory canal, carotid canal and mastoid. CT scans demonstrated extensive infiltration and permeative destruction of the skull base (Fig. 4). On MRI the tumor was markedly heterogeneous with multiple hyperintensive areas on non-contrast T1-weighted images consistent with subacute hemorrhage (Fig. 5). *Hemangiopericytoma* was a localized mass located in the jugular foramen with limited invasion of the medial mastoid (Fig. 6). *Endolymphatic sac carcinoma* occupied the jugular foramen and the retrolabirynthine portion of the petrous bone. Limited

Table 1 – Clinical details and summary of radiologic features of presented jugular foramen tumors.

Tumor/no Modality/no No/Sex (age Y)	Tumor epicenter/ Prominent extension	Growth vector/pattern	CT – bone changes	Other CT features	MRI T1 weighted T1 + contrast	MRI T2 weighted	Angiography
Schwannoma/4 CT/4, MRI/4, DSA/4 3/M (18, 29, 36) 1/F (34)	Petrous bone epicenter: JF/ Parapharyngeal space	Expansile growth along CN; ICA displacement;	No bone infiltration; Cortex preservation, Smooth JF enlargement		Low SI, Strong, irregular CE	High SI, Lobulated, Heterogeneous	Hypovascular
Meningioma/2 CT/2, MRI/2, DSA/2 2/F (36, 50)	Petrous bone epicenter: JF/ CPA	Multidirectional growth/ INCR: dural planes; ICA encasement;	Diffuse infiltration, Cortex erosion, Limited JF widening, Preserved trabecular architecture	Hyperostosis, Calcifications	Low SI; Strong, uniform CE, Dural tails	Low SI;	1/2 hypovascular, 1/2 hypervascular: intense blush
Ependymoma/1 CT, MRI, DSA 1/F (33)	Petrous bone epicenter: JF/ Large CPA extension	Multidirectional growth extensive	Diffuse infiltration, Cortex erosion, Irregular JF enlargement Extensive permeative destruction	Intratumoral bony trabeculae	Heterogeneous, Hyperintense foci, Strong, irregular CE	Heterogeneous, Flow voids, Hemosiderin	Hypervascular, intense blush
Hemangiopericytoma/1 CT, DSA 1/F (67)	Petrous bone epicenter: JF	Limited lateral: mastoid	Limited infiltration, Cortex erosion, Irregular JF enlargement, Permeative destruction;		Not performed	Not performed	Hypervascular, intense blush
ELST/1 CT, MR 1/F (27)	Petrous bone epicenter: retro-labyrinthine	Limited in all directions	Limited infiltration, Cortex erosion, Permeative/geographic destruction	Bony fragments, Posterior rim calcification	Heterogeneous, Hyperintense areas, Osseous foci, Strong, irregular CE	Heterogeneous, Hyperintense areas, Osseous foci,	Not available
Metastasis/2 CT/2, MRI/1 2/F (45, 57)	Petrous bone epicenter: CC and JF	Multidirectional, Circumferential/ ICA encasement + stenosis	Lytic destruction, Loss of cortex and trabeculae, Irregular JF enlargement		Low SI, Homogeneous, Uniform CE	Intermediate SI, Homogeneous,	Not performed
Paraganglioma/10 CT/10,MRI/10,DSA/10 7/F (52–61) 3/M (46–58)	Petrous bone epicenter: JF	Predominant superolateral: middle ear/ ICA displacement	Limited infiltration, Cortex erosion, Irregular JF enlargement, Permeative destruction		Low SI, Strong, irregular CE	Intermediate SI, Flow voids,	Hypervascular, intense blush

CC – carotid canal; CE – contrast enhancement; CN – cranial nerves; CPA – cerebelloopontine angle; ELST – endolymphatic sac tumor; ICA – internal carotid artery; INCR – intracranial; JF – jugular foramen; SI – signal intensity

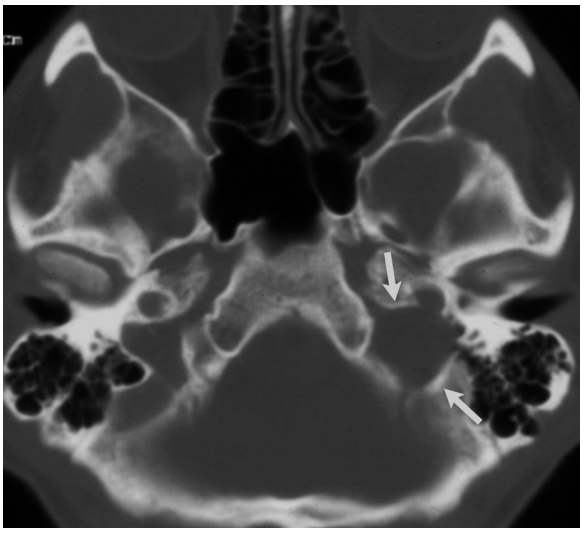

Fig. 1 – Schwannoma. Axial bone-window CT scan shows smooth widening of the jugular foramen with preserved rim of cortical bone (arrows).

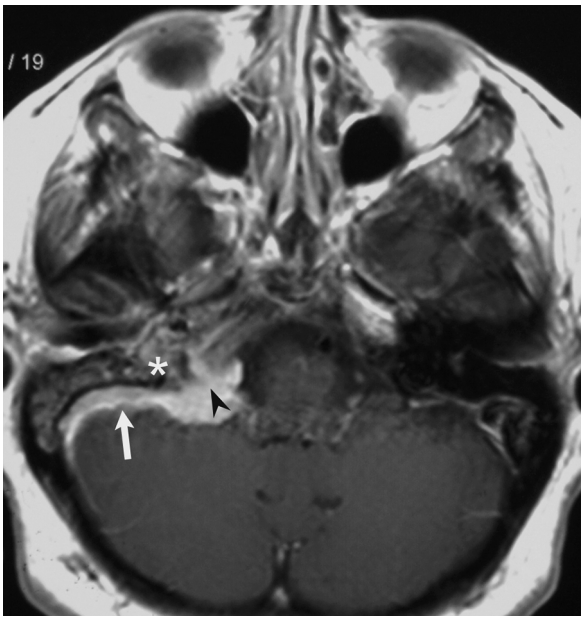

Fig. 3 – Meningioma. Axial contrast-enhanced T1-weighted image shows prominent dural enhancement (arrow) and hyperostosis (asterisk), without significant widening of the jugular foramen (arrowhead).

extensions to the posterior fossa, the middle ear cavity and medial mastoid were observed (Fig. 7). Both *metastases* occupied the jugular foramen and the horizontal segment of the carotid canal, and caused encroachment and stenosis of the ICA below the skull base. Involvement of the posterior cranial fossa occurred in one case. *Paragangliomas* (10) were centered in the jugular foramen (Fig. 8). All tumors showed superolateral spread involving the middle ear cavity (10/10), whereas involvement of the posterior cranial fossa (1/10) and carotid canal (2/10) were less common. Inferior spread was limited, with no encasement of the ICA.

4. Discussion

Clinical presentation of jugular foramen tumors typically corresponds to encroachment of vital structures located within the jugular foramen or in adjacent anatomical areas. Most commonly patients present with the pulsatile tinnitus, hearing loss and/or a combination of cranial nerve palsies (VII, IX, X, XI, XII) [1,3]. Although paragangliomas are the most frequent jugular foramen tumors, the differential diagnostic

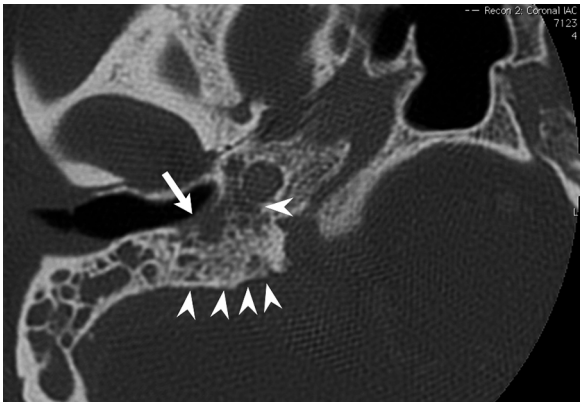

Fig. 2 – Meningioma. Axial bone window CT scan shows petrous bone infiltration with predominant sclerotic changes and preserved trabecular structure (arrowheads). Invasion of the middle ear cavity (white arrow).

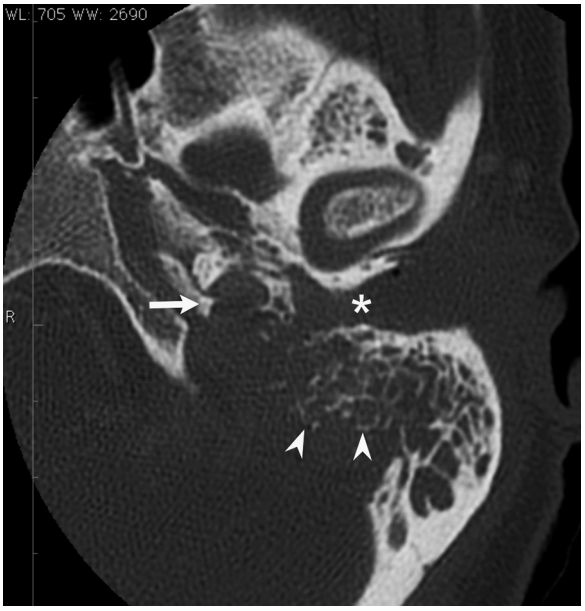

Fig. 4 – Ependymoma. Axial bone window CT scan shows irregular enlargement of the jugular foramen with skull base infiltration and intratumoral trabeculae (arrowheads). Invasion of the carotid canal (arrow), tympanic cavity and external auditory canal (asterisk) is present.

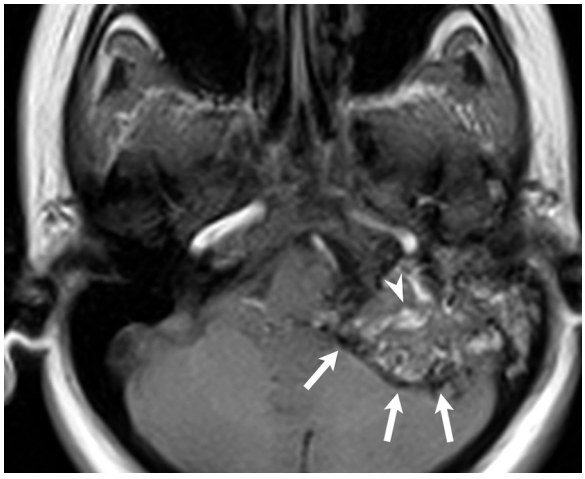

Fig. 5 – Ependymoma. Axial unenhanced T1-weighted image demonstrates large tumor with areas of high signal intensity (arrowhead) and hypointensive rim suggestive for hemosiderin (arrows).

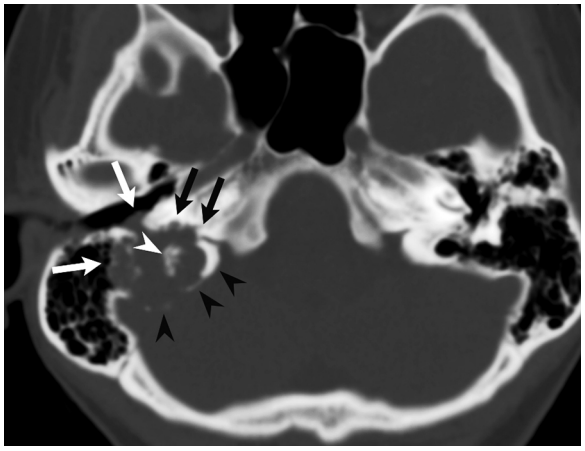

Fig. 7 – Endolymphatic sac tumor. Axial bone window CT scan shows geographic bone destruction (black arrows) and structures of bone density within tumor mass (arrowhead) and along its posterior margin (black arrowheads). Limited invasion of the tympanic cavity (white arrows).

list of possible primary and secondary lesions is extensive and includes schwannoma, meningioma, chordoma, chondrosarcoma, endolymphatic sac tumor, giant cell tumor, adenocarcinoma, plasmocytoma, histiocytosis, metastasis [1–5]. These lesions may have identical clinical manifestation, making the correct diagnosis on the basis on clinical symptoms impossible. However, due to different histopathological nature they require different therapeutic approach. As preoperative radiological diagnosis has important therapeutic implications a radiologist should be familiar with important differentiating features and similarities in imaging appearances of rare jugular foramen tumors in comparison to most common lesions of this region.

Schwannomas of the jugular foramen show expansile growth and widening of the jugular foramen is associated with pressure erosion. Involvement of the skull base is limited, because, unlike paragangliomas and meningiomas, they do not infiltrate the surrounding bone. Schwannomas may be classified into four groups: A – primarily intracranial tumors with enlargement of the jugular foramen; B – primarily jugular foramen tumors; C – primarily extracranial tumors with involvement of the jugular foramen; D – tumor with intra- and extracranial part and ''dumbbell-shaped'' appearance [6]. In our study type C growth pattern occurred in 3 patients with large extracranial component, whereas ''dumbbell-shaped'' tumor was stated in 1 patient. The growth vectors lead along the IX-XI cranial nerves and the superior aspect of the tumor is

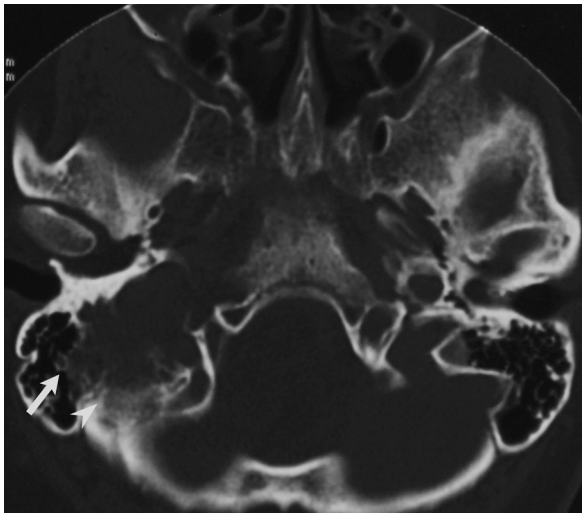

Fig. 6 – Hemangiopericytoma. Axial bone window CT scan shows irregular jugular foramen widening with erosion of cortical margins and permeative growth pattern (arrowhead). Involvement of the medial mastoid (arrow).

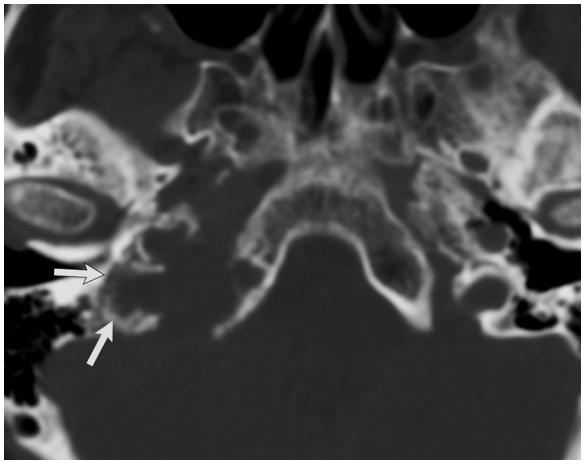

Fig. 8 – Paraganglioma. Axial bone-window CT scan demonstrates limited bone infiltration, permeative erosion of cortical margins (arrows) and irregular widening of the jugular foramen.

generally directed superomedially toward the brainstem [7]. Characteristic CT features of the jugular foramen affected by schwannoma are pressure expansion and scalloping with smooth and well demarcated cortical margins, in contrast to indistinct border and permeative erosion of adjacent bone suggestive for paraganglioma [2,5,8,9]. On MRI we observed a lobulated appearance and heterogeneous contrast enhancement in all cases of schwannoma. Cystic component may be seen in up to 25% of cases, while calcifications or hemorrhage are uncommon [1,2,9]. Flow voids have not been reported. Typically the cervical ICA is anteromedially displaced, but not surrounded by the tumor mass [9]. Angiographic findings in schwannomas are quite different from the pattern seen with paragangliomas, as schwannomas are typically hypovascular.

Jugular foramen meningiomas are characterized by centrifugal type of growth, which takes place in all directions: the middle ear cavity, CPA, jugular tubercle, clivus, below the skull base [10]. We observed predominant superior growth into the posterior cranial fossa. Intracranial spread follows dural planes. It may result in less common soft tissue mass with broad dural attachment or may show ''en plaque'' appearance, without distinct intracranial mass, which is characteristic for primary jugular foramen meningiomas [2,5,7]. Extensive extracranial growth, observed in one of our patients, is uncommon, as less than 1% of intracranial meningiomas involve the lower part of the parapharyngeal space [8,11]. Chen et al. [11] reported a case of meningioma involving jugular foramen and cerebellopontine angle, with extension to the parapharyngeal space and encasement of the common carotid artery bifurcation. Encroachment of cervical ICA was reported also by Vrionis et al. [12]. We observed this feature in both meningiomas, in contrast to paragangliomas and schwannomas. In primary jugular foramen meningiomas bone destruction is less pronounced, than in paragangliomas. Meningiomas show wide infiltration of the diploic spaces of the skull base, with relative preservation of the internal trabecular architecture [5]. Irregular erosion of normal cortical margins may be observed, but widening of the jugular foramen is rather limited [1,2]. MacDonald et al. [7] found this CT appearance in all patients in his study and referred to this growth pattern as ''permeative-sclerotic'' in contrast to ''permeative-destructive'' spread seen in glomus jugulare tumors and expanded, well corticated margins in schwannomas. Calcifications and hyperostosis are rare, but characteristic for meningiomas [1,2,4]. On MRI the signal intensity of meningiomas may be low on T2-weighted images, depending on the histopathology and the presence of calcifications [1,2,4,5]. ''Dural tails'' is a typical finding on post-contrast images. Further features useful for differential diagnosis include lack of flow-voids characteristic for paragangliomas and no cystic components, which may occur in schwannomas [4,10]. Angiographic appearance of meningiomas varies from minimal to high vascularity, which depends probably on the histological composition [2]. Most tumors lack early blush characteristic for paragangliomas [4,12]. However, vascular meningiomas may occur [11]. We observed rich tumor vasculature in one case of meningioma.

Cerebellopontine angle ependymomas are rare and tumor involving the jugular foramen has not been reported [13–15]. Due to unique location differential radiological diagnosis in ependymoma of the CPA and jugular foramen is a challenge. Tumor is characterized by large posterior fossa extension. However, although not typical, extensive intracranial growth with CPA involvement may occur in jugular bulb paragangliomas [16,17]. Within the skull base ependymoma shows infiltrative growth pattern, with diffuse skull base involvement and extensive centrifugal spread. In contrast to meningiomas, bone-window CT demonstrates extensive permeative bone destruction, with enlargement of the jugular foramen and multiple intratumoral bony fragments. No hyperostosis or sclerotic changes have been observed. On MRI ependymoma is markedly heterogeneous in contrast to meningiomas. Despite the presence of signal voids, speckled tumor appearance with multiple hyperintensive foci on T1-weighted images and rim of hemosiderin is quite different from the ''salt and pepper'' sign characteristic for paragangliomas. MRI findings of ependymoma in our study resembled findings of CPA ependymomas, including irregular margins, heterogeneity, low signal intensity on T1-weighted images and low to high signal intensity on T2-weighted images, hemorrhage, heterogeneous contrast enhancement, limited peritumoral edema [18]. Angiographic findings in ependymoma imitated jugular foramen paraganglioma.

The presentation of hemangiopericytoma in the jugular foramen is very rare, with only 3 cases described in the literature [3,19,20]. This tumor has an appearance of a localized mass. Infiltration of the skull base and invasion of surrounding anatomical areas are limited. Hemangiopericytoma is characterized by bony changes similar to observed in paragangliomas. Angiography shows hypervascular tumor with blush characteristic for paraganglioma. Comacchio et al. [20] observed on CT scans wide osteolytic process. In other reports radiographic findings, including angiography, were consistent with paraganglioma [3,19]. While subtotal resection or radiation therapy might be considered in some paragangliomas, total removal is the treatment of choice for hemangiopericytomas, due to the incidence of malignant disease approaching 50% [19]. As incorrect preoperative diagnosis of glomus jugulare tumor may results in suboptimal treatment, some authors suggest additional diagnostic work-up including pre-treatment biopsy [3,19].

Endolymphatic sac tumor (ELST) is primarily centered in the retrolabirynthine portion of the petrous bone, between the internal auditory canal and the sigmoid sinus, in the area of the vestibular aqueduct. This is appreciable even in advanced tumors, in contrast to paragangliomas, with predominant infralabirynthine location [21,22]. Tumor grows in all directions invading supra- and infralabyrinthine regions, tympanic cavity, medial mastoid, posterior fossa. Intracranial spread may lead to dural transgression, which was not observed in our study [22]. Unlike schwannomas, ELST shows irregular bone destruction with geographic and/or permeative bony margins. Infiltration of the skull base is limited in contrast to meningiomas and ependymomas. Tumor mass is characterized by the presence of multiple intratumoral bony fragments of reticulated, spiculated or stippled appearance [2,22]. Characteristic finding for ELST is posterior rim calcification, which may correspond to posterior face of the petrous bone displaced posteriorly by enlarging tumor [21]. Unlike schwannomas and

meningiomas these tumors are markedly heterogeneous on MRI. T1-weights images show hyperintense areas suggestive for subacute hemorrhage and/or proteinaceous cysts, as well as strong, heterogeneous enhancement after contrast administration. Lo et al. [22] reported one case of nearly homogenous tumor with a short segment of dural enhancement resembling meningioma. ''Dural tails'' were not observed in other cases of ELST [21,22]. Angiography shows hypervascular tumor imitating paraganglioma [2,4].

Metastases involving the jugular foramen most commonly derive from primary malignancies of the head and neck, breast, lung, kidney and prostate [23]. The results of the recent study show that the incidence of nasopharyngeal cancer metastases in the skull base is low and that they occur most frequently in the jugular foramen and the horizontal segment of the ICA [24]. Metastatic tumors in both our patients occupied the jugular foramen, the ICA canal and petrous apex, with limited posterior fossa invasion in one case. Although extensive involvement of the ICA canal and petrous apex may be seen in paragangliomas, this growth direction without posterolateral spread to the middle ear and mastoid is not typical for this tumor. Characteristic radiologic features of malignant lesion are aggressive, circumferential growth, without respect of fascial and osseous borders, as well as encroachment and thinning of the ICA below the skull base [1,9]. CT scans typically demonstrate lytic destruction of the petrous bone, with irregular contour of bony margins [1,5,9]. Some metastases may show sclerotic or mixed osseous changes, particularly in prostate cancer [4,25]. On MRI metastatic tumors usually are hypointense on T1-weighted images and lack high signal on T2-weighted images [5]. Strong contrast enhancement and tubular signal voids may be seen in vascular lesions, such as those deriving from the renal and thyroid cancer [5,26,27]. Radiological findings in such cases may be difficult to distinguish from glomus jugulare tumors [1]. However, skull-base metastases usually occur in advanced stage of the disease, when many patients already have disseminated and recognized cancer [2,4,25].

Both CT and MRI are widely used for the evaluation of lesions at the jugular foramen.

High-resolution CT of bone window is useful as a first step in the differential diagnosis of paragangliomas, schwannomas and meningiomas, accounting for over 90% of jugular foramen masses, as these three tumors cause different bony changes. MRI gives additional information, as it allows more comprehensive analysis of tumor nature and shows some important tissue characteristics. Best evaluation of jugular foramen lesions requires high-quality cross-sectional imaging with both modalities to obtain sufficient information for correct differential diagnosis and to fully answer all preoperative questions [4,5,9]. Angiography demonstrates tumor vascular composition and aids the pathological diagnosis, but its main goal is preoperative embolization in patients, who may benefit from this procedure. Although at the jugular foramen angiographic appearance of highly vascular tumor with intensive blush is most commonly seen in paragangliomas, it may also be observed in rare tumors, like meningioma, hemangiopericytoma, ependymoma, endolymphatic sac tumor or some metastases [3,11,26,27].

5. Conclusion

A combination of certain radiological features including tumor epicenter, growth vectors, skull base infiltration, bony changes and tumor morphology help establish correct preoperative diagnosis and differentiate less common jugular foramen tumors, such as schwannoma, meningioma, ependymoma, ELST or metastasis, from most common paragangliomas

The diagnosis of jugular bulb paraganglioma can be made with high probability when typical imaging findings are present. However, hemangiopericytoma may have similar radiological appearance and must be considered in differential diagnosis.

Conflict of interest

Authors declare no conflict of interest.

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