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Case Report

Paraganglioma of the cauda equina region

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Abstract

BACKGROUND CONTEXT: Cauda equina paragangliomas (CEPs) are rare neuroendocrine tumors. The difficulty in differential diagnosis with other tumors of this region may be misleading for surgical planning and prognostic expectations.

PURPOSE: To report on a rare case of CEP and review the most current information regarding the diagnosis, treatment options, and outcomes.

STUDY DESIGN: Case report and literature review.

PATIENT SAMPLE: One patient affected by CEP.

METHODS: We report on a 33-year-old woman with a 2-month history of worsening low back pain, aggravated by sitting, bending, and coughing. Neurological examination revealed normal power and muscular tone, no sensory or sphincter abnormality, and normal reflex. Magnetic resonance imaging of the lumbar spine demonstrated an intradural extramedullary lesion at L3, with homogeneous contrast enhancement and hypointense punctate foci. The patient underwent an L3 laminectomy and tumor removal. Relevant articles covering CEPs from 1970 to the present were reviewed.

RESULTS: The histopathological examinations described paraganglioma features. The postoperative course was uneventful, and all the symptoms resolved, with no tumor recurrence after 3 years' follow-up.

CONCLUSIONS: Cauda equina paragangliomas are rare, benign, and slow-growing tumors. Except for its secreting tumor characteristics, preoperative CEP diagnosis is very difficult. Magnetic resonance imaging is important and may suggest specific radiological features for these tumors; however, these are only relative, and it is rare that diagnosis is made before surgery. Diagnosis is established by histological examination and electron microscopy, and immunohistochemical techniques must be used to achieve a correct diagnosis. Cauda equina paragangliomas are well-encapsulated tumors that may be cured by surgery alone, whereas radiotherapy is reserved for incompletely resected tumors. Overall, prolonged postoperative observation is mandatory because of the slow tumor evolution and the possibility of tumor relapse even up to 30 years after surgery.

Keywords: Paraganglioma; Cauda equina; Filum terminalis; Recurrence; Surgery; Radiotherapy

Background

Paragangliomas (PGLs) are rare and well-recognized neuroendocrine tumors that derive from the neuroepithelial cell group called paraganglia [1–3].

Paragangliomas may occur throughout the body, but most commonly occur in the head and neck regions (more than 90% of cases) mainly from the carotid bodies (chemodectomas) and the jugular glomus. However, they have been described in other locations, such as the urethra, liver, larynx,

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duodenum or retroperitoneum, para-aortic body, and orbit, rarely involving the central nervous system [1,4,5].

Cauda equina paragangliomas (CEPs) are exceptional and represent approximately 3.5% to 4% of all neoplasms in this region, considering that they have been steadily reported approximately four to eight times per year worldwide [2,6,7]. The difficulty in differential diagnosis with other tumors of this region, namely ependymomas and neurinomas, may be misleading for surgical planning and prognostic expectations.

We report on a new case of CEP, reviewing all the pertinent literature.

Case report

A 33-year-old woman came to our facility with a 2-month history of worsening low back pain that had a nocturnal predominance and an intermittent tingling sensation in the inguinal-crural area.

Within the past 10 days, the pain had intensified and was aggravated by sitting, bending, and coughing. Neurological examination revealed normal power and muscular tone, no sensory or sphincter abnormality, and normal reflex.

Magnetic resonance imaging of the lumbar spine demonstrated an intradural extramedullary lesion at L3 (Fig. 1). The T2-weighted (T2-w) images revealed a lobulated lesion with heterogeneous signal intensity (Fig. 1, Top Left). Low signal intensity regions were detected within the lesion, suggesting hemorrhage, and serpiginous vascular structures were detected extending from the conus medullaris to the tumor. Contrast-enhanced T1-weighted (T1-w) imaging demonstrated homogeneous enhancement with hypointense punctate foci (Fig. 1, Top Left and Top Right), which is associated with a serpiginous enhancing vascular structure.

The patient underwent an L3 laminectomy, and the exposed dural sac was opened at the midline. The tumor nodule was visualized deep to the cauda equina roots.

The tumor appeared to be a well-encapsulated light grayish-red and glossy lesion covered by nerve roots; it originated from the cauda equina nerve roots. The tumor was circumferentially dissected from the nerve roots to facilitate removal. The lesion was firmly attached over its ventral surface to two nerve roots, one of which was cautiously dissected, whereas the other was coagulated and cut (Fig. 2). The tumor was eventually removed en bloc. The patient's hemodynamics remained stable during the tumor manipulation. The dura was then closed in a watertight fashion, and the wound was closed in layers.

Pathological studies confirmed the PGL diagnosis (Fig. 3). The postoperative course was uneventful, and all the symptoms resolved. The patient was discharged 5 days after the surgical procedure. There is no sign of recurrence after 3 years of follow-up.

Discussion

By an extensive literature review, we found 254 cases reported in 44 years, since 1970. In accordance with

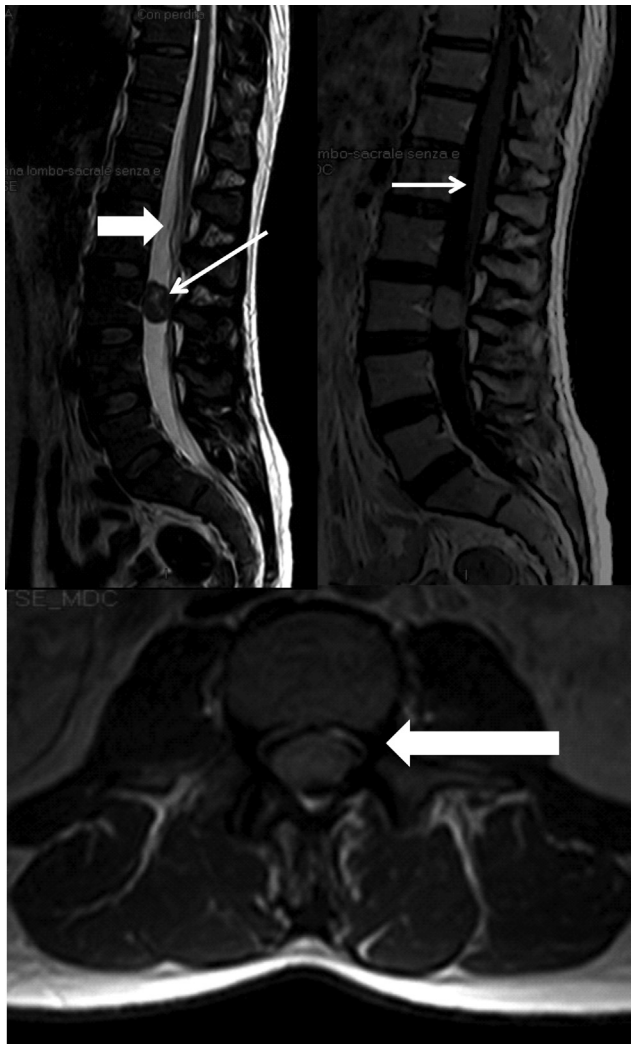

Fig. 1. (Top Left) The sagittal T2-weighted magnetic resonance (MR) image revealed a lobulated lesion with heterogeneous signal intensity, regions of low signal intensity within the lesion, suggesting hemorrhage (thin white arrow); serpiginous vascular structure extending from the conus medullaris to the tumor was also visible (thick white arrow). (Top Right) Sagittal contrast-enhanced T1-weighted MR image demonstrated homogeneous enhancement with hypointense punctate foci and also the serpiginous enhancing vascular structure (white arrow). (Bottom) The axial contrast-enhanced T1-weighted MR image showed a well-defined spherical-enhanced tissue filling almost the entire spinal canal, and the punctate structure (thick white arrow) indicates a vessel appreciated on resection of tumor.

Gelabert-Gonzàlez [2], we have excluded from the analysis those cases with noncomparable information, reaching a total amount of 234 cases for discussion [1–5,8–41].

Etiopathological considerations

Paragangliomas are rare and usually slow-growing neuroendocrine tumors that arise from the neuroepithelial cell group called paraganglia. These paraganglia are subdivided into two groups: one is formed by adrenal medulla and the other is formed by the extra-adrenal paraganglia [2]. Although pheochromocytoma is a term reserved for tumors arising from the adrenal medulla, PGL characterizes tumors

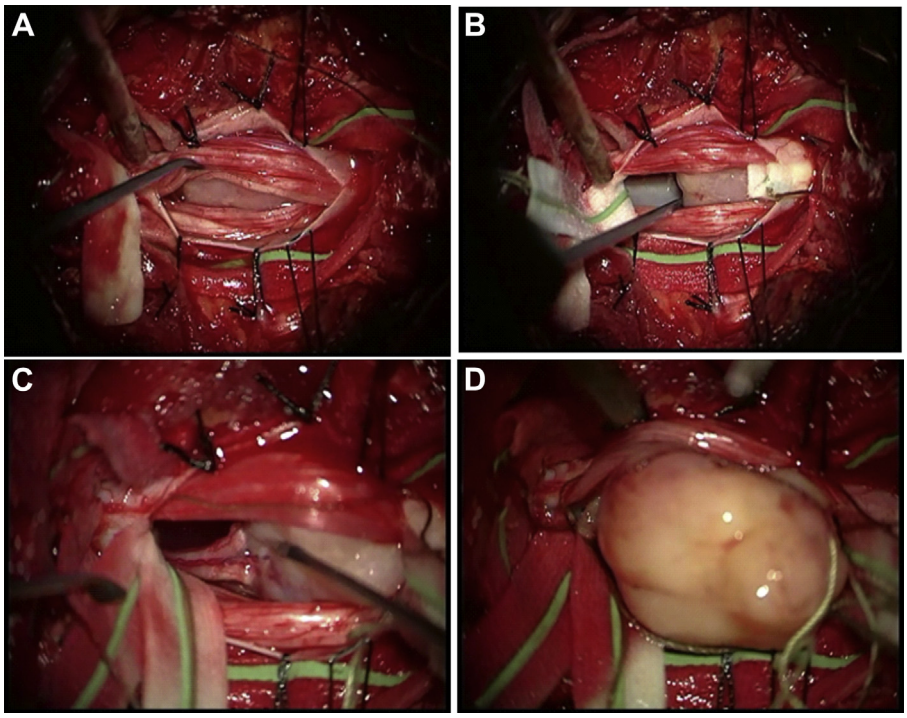

Fig. 2. Intraoperative images of cauda equina paraganglioma. (A) Cauda equina paraganglioma (CEP) under nerve roots after opening of the dural sac. (B) CEP dissected from the nerves roots. (C) CEP firmly attached over its ventral surface to two nerve roots. (D) CEP after resection.

deriving from extra-adrenal paraganglia, and chemodectoma is limited for PGLs deriving from the carotid body and related paraganglia, which are usually parasympathetic [6].

These tumors retain two fundamental biochemical characteristics: they can synthesize and store biogenic amines belonging to the amine precursor uptake decarboxylase

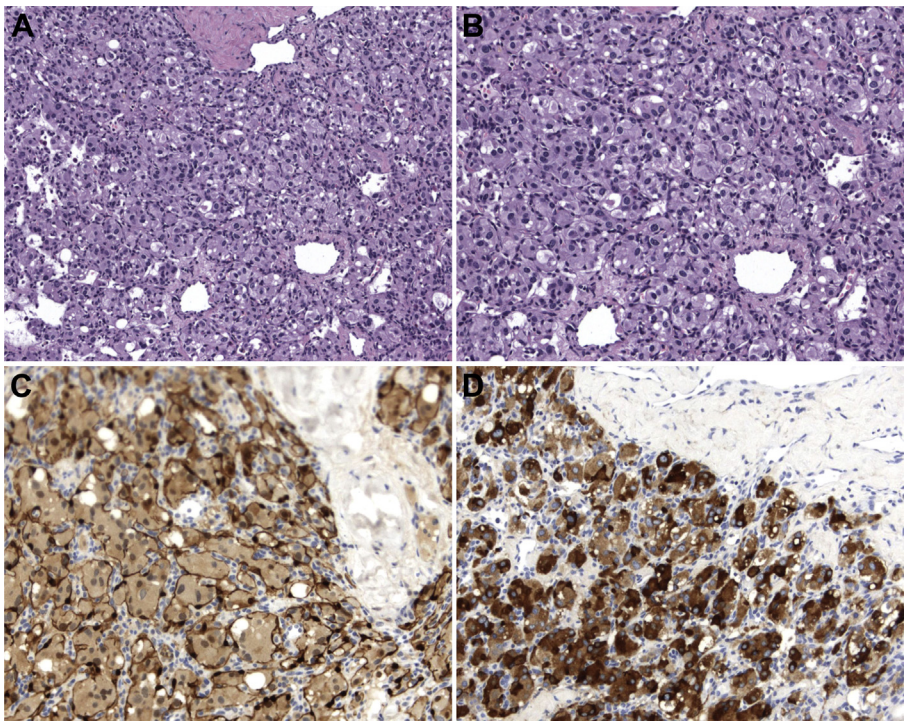

Fig. 3. (A) Hematoxylin and eosin 200×, nest of tumor cell, consisting of round small tumor cell, separated by a fine vascular network, known as a "Zellballen" pattern. (B) Hematoxylin and eosin 400×, the reticulin staining highlighted this pattern; tumor cell showed a pseudorosette pattern, the nuclei were uniformly round to oval. (C) 200×, immunohistochemical staining was positive for synaptophysin and chromogranin A. (D) 200×, the expression of S-100 showed the existence of sustentacular cells. The tumor cells were negative for glial fibrillary acidic protein and epithelial membrane antigen.

system, and they can produce possibly secreted peptide hormones (adrenaline, noradrenaline, and dopamine) [2,3].

Paragangliomas with en extraspinal localization may be multicentric in 3.8% to 10% of sporadic cases and have a higher incidence near 25% to 35% in subjects with a positive family history. They may also form part of the multiple endocrine neoplasia type 2 and Von Hippel Lindau disease [6,36].

Spinal localization of these tumors is unusual; the estimated annual incidence is reportedly 0.07 per 100,000 in the general population [36]. Cauda equina paragangliomas represent approximately 3.5% to 4% of all neoplasms in this region [2,6,7].

Spinal PGLs are believed to originate from sympathetic neurons placed in the thoracic and lumbar horns of the spinal cord, for which the path to the efferent sympathetic chain is represented by spinal communicating branches [2,3,8]. Another theory is that tumors may arise from heterotopic neurons that lie along these proximal branches. In the lumbar region, they may arise from paraganglia that are located in the cauda equina. Because ependymal cell participation in their development cannot be ruled out, some authors speculate that some diffuse neuroendocrine system constituents can be the result of a local differentiation from the tissue that was not derived from the neural crest [4].

Epidemiology

The first description of CEP was published in 1972 by Lerman et al. [9]; however, 2 years prior, Miller and Torack [8] had already described a case of "secretory ependymoma of the filum terminale."

Patients ranged in age from 9 to 77 years (mean, 46.4±13.5 years), with peak incidence during the fourth/fifth decade. There was a male predominance (male:female ratio is 1.4:1); of the series of patients, there were 137 males ranging in age from 13 to 77 with a mean age of 46.0±12.8 years and 97 females ranging in age from 9 to 74 with a mean age of 48.2±14.4 years (Table 1).

Clinical presentation

The most frequent initial symptom was low back pain, which was present in 43.1%; low back pain and sciatica were reported in 23.9% and unilateral sciatica in 8.1%. Motor deficit was detected in 6.8% of cases and sensory deficits in 2.5%, whereas sphincter and erectile dysfunctions were reported, respectively, in 2.9% and 1.2% of the patients. Clinical intracranial hypertension symptoms were observed in 2.9% of cases. Commonly, diagnosis was delayed by more than 1 year after presentation (range, 1–7 years), which reflects the nonspecific nature of the symptoms [14]. It is interesting that although the mass lesion may occupy the entire spinal canal diameter, it is unusual for the lesion to cause a cauda equina syndrome until very late [14,16,27,41].

Table 1
Summary of clinical characteristic

Epidemiology	
Patient population	234
Sex (M/F)	137/97
Mean age at clinical onset	46.4 y
Clinical presentation	**%**
Low back pain	43.1
Low back pain and sciatica	23.9
Unilateral sciatica	8.1
Motor deficit	6.8
Cauda equina syndrome	2.9
Raised ICP symptoms	2.9
Sphincter dysfunction	2.9
Sensory deficits	2.5
Erectile dysfunction	1.2
Hemorrhagic manifestations	0.8

M, male; F, female; ICP, intracranial pressure.

In extremely rare cases, the clinical onset may be represented by acute severe neurological deterioration related to iatrogenic maneuvers: Djindjian et al. [15] reported one case of sudden paraplegia after sacral medication infusion for low back pain treatment, and Pikis et al. [40] reported the case of a patient with acute cauda equina syndrome after spinal epidural steroid injection.

Systemic manifestations, which are frequently associated with pheochromocytomas because of catecholamine release, are rarely described in CEP because of a supposed inability of these tumors to secrete hormones into the bloodstream or the inefficacy of the released substances to provoke symptoms [6].

These tumors are often described as high vascular lesions; however, hemorrhagic manifestations, namely subarachnoid hemorrhage, represented the onset of clinical findings in only two patients [6,28,34].

Radiological features and differential diagnosis

The gold standard for diagnosis is magnetic resonance imaging, although a correct diagnosis may pose some problems because the neuroradiological features of the lesion are generally nonspecific.

The lesion on T1-w sequences is usually hypo- or isointense to the conus medullaris; however, it is hyperintense on T2-w sequences, and sometimes heterogeneous signal intensity has been observed [20].

Hypointense tumor margins on T2-w sequences and gradient echo imaging may indicate paramagnetic effects from hemosiderin or ferritin because of previous hemorrhages [4,19,20,25].

Paragangliomas are hypervascular lesions that produce punctuate areas of flow void dispersed in a matrix of increased signal intensity. Intralesional punctate and linear low signal and multiple signal voids capping the tumor were likely to be findings of dilated vessels. On T2-w sequences, a salt-and-pepper appearance is distinctive of head

and neck PGLs [42]. Contrast-enhanced T1-w sequences demonstrated marked enhancement [19], and serpiginous flow void around the tumor was a frequent feature, suggesting that dilated vessels or congested veins attributed to the hypervascularity [26,30].

High signal intensity was observed in the cerebro-spinal fluid (CSF) beneath the tumor, probably secondary to CSF circulation blockage, CSF protein elevation, or CSF pulsation reduction [21]. According to the literature, cystic areas are reportedly located in one lesion pole [4].

When performed, selective spinal angiography may help the diagnosis because it demonstrates a highly vascular mass in the early arterial phase. The vascular blush becomes homogeneous in the late arterial phase. Preoperative embolization of the vessels feeding the tumor may be performed to minimize intraoperative bleeding [13].

Currently, scintigraphy with I-131 metaiodobenzylguanidine is a functional imaging method for adrenal and extra-adrenal PGL localization. Although this method provides a high level of sensitivity and specificity, it is also inconvenient because of elevated radiation exposure, limited spatial resolution, and failure to identify nonsecreting PGLs, which are the most of those situated in the lumbar region [2].

Preoperative CEP diagnosis is almost impossible, except in the presence of systemic manifestation with the possibility to detect high urinary biogenic amines or their metabolites [4,13].

The other intradural extramedullary lesions that must be considered in the differential diagnosis are myxopapillary ependymomas, ependymomas, schwannomas, meningiomas, hemangioblastomas, metastatic tumors, epidermoids, and lipomas [20,24]. However, these characteristics are only relative, and diagnosis is rarely made before surgery.

Histopathological findings

The typical appearance on histopathological examination is a uniform population of small, polyhedral, round, columnar, or cylindrical to cuboidal cells, which form a nest or ''zellballen'' that is circumscribed by a dense connective tissue containing a delicate and extensive network of endothelial-lined vessels [31,32].

The predominant cell type is the chief cell, which has abundant eosinophilic granular cytoplasm; the second cell type is the sustentacular or supporting cell. The nuclei are uniformly round to oval and generally contained nucleoli.

Paraganglioma that has lost the normal paraganglionic structure has been associated with a more aggressive or malignant behavior; the paucity or the absence of sustentacular cells is indicative of an aggressive or malignant nature [2].

This lesion demonstrates an assortment of histological growth patterns, such as alveolar, spindle cell, oncocytic, gangliocytic, papillary, and ribbing; pseudorosette formation is a frequent feature [6].

Immunohistochemical staining is most commonly used for differential diagnosis; the chief cells are neuroendocrine in origin and positive for neuron-specific enolase, chromogranin, and synaptophysin [2]. Generally, tumor cells are negative for epithelial membrane antigen and glial fibrillary acidic protein [42]. The characteristic finding is that sustentacular cells are sensitively and reliably expressed by S-100 protein [27].

Ultrastructurally, the cytoplasm demonstrates electron-dense neurosecretory granules that are 150 to 400 nm on size [2].

Biochemical analysis demonstrates the presence of somatostatin, serotonin, noradrenaline, adrenaline, dopamine, and homovanillic acid [10]. Biochemical and immunohistochemical analyses of the spinal PGL revealed that 5-hydroxytryptamine was present in PGL at higher levels than catecholamines [39]. Sonneland et al. [14] revealed that up to 75% of their tumors demonstrated 5-hydroxytryptamine immunoreactivity.

However, functional neurosecretory granules are very rare in CEPs [11]. The lack of paroxysmal hyperadrenergic states in spinal PGL may have two reasons: the inability of tumor cells to secrete stored substances or the inability of these substances to provoke a clinical syndrome [43].

Immunohistochemical study allows for distinguishing PGLs from ependymomas (positive for glial fibrillary acidic protein and mucin, with different pattern of S-100 protein), epithelioid schwannomas (diffuse positivity for S-100), and meningiomas (consistent reactivity both for epithelial membrane antigen and vimentin) [2].

Surgical considerations and biological behavior

The preoperative workup for differential diagnosis, including appropriate neuroradiological and laboratory examinations, plays a key role in the surgical planning. This raises important considerations for surgical management to avoid possible intraoperative complications related to CEP characteristics, namely hemorrhagic and systemic hyperadrenergic manifestations.

Excess catecholamine production with systemic complications has been reported in only four cases. One case had flush-like attacks without paroxysmal hypertension but with abnormal urine noradrenaline concentrations [11]. The other three cases presented with hyperadrenergic symptoms either immediately preoperatively after positioning in the knee-chest position or intraoperatively while the tumor was manipulated [2,4,37]. All the symptoms subsided when the vascular pedicle was clamped. Nevertheless, hemodynamic liability with vasomotor amine syndrome poses an important comorbidity for patients under general anesthesia. Tumor manipulation can trigger a hypertensive crisis, which is extremely important in patients with underlying coronary artery disease and limited cardiac reserve [8].

Catecholamine metabolites vanillylmandelic acid and metanephrine can act as tumor markers; levels of these metabolites may be obtained as a preinterventional baseline.

Cauda equina paragangliomas are highly vascular lesions, which are easily identified on selective spinal

angiography in the early arterial phase. Preoperative tumor embolization has been used in some cases to reduce intraoperative bleeding and catecholamine load [2,6].

Although small intratumoral hemorrhage was often noted, massive bleeding within and outside the lesion is rare. Demirçivi Ozer et al. [1] reported one case of postoperative hemorrhage at the surgical site in which the patient developed paraplegia, whereas Matsumoto et al. [38] described a hematoma and pulsating bleeding at the lateral aspect of the tumor, and this patient presented with headache and meningeal irritation.

Cauda equina paragangliomas are usually soft, well-encapsulated, and well-circumscribed masses, and total removal should be the goal of surgery. Whenever possible, the tumor should be resected in one piece with an intact capsule to prevent subarachnoid dissemination [6].

Using the World Health Organization grading system, CEPs are classified "*Tumors of Neuro-Epithelial Tissue*" in the subgroup "*Neuronal and Mixed Neuronal-Glial Tumors*" as Grade 1 [44], and an aggressive behavior occurs in less than 1% of tumors [27,36].

The clinical behavior depends mainly on encapsulation and the extent of resection [6]. Overall, the prognosis is good because a complete removal can be achieved in most cases. Indeed, when a subtotal excision is performed for local invasion, local recurrence is not infrequently observed, and even distant metastatic spread has been described [14].

According to cases review, all the patients underwent surgery; in 193 cases (82.5%), a gross total resection (GTR) was achieved, a subtotal resection (STR) was performed in 18 cases (7.7%), and two biopsies; radiotherapy was undertaken in 25 cases (8 GTR cases, 15 STR cases, and two biopsies). No mention of the extent of removal was reported in 21 cases.

In 34.7% of cases, the tumor was attached to a nerve root or the cauda equina, in 30% it was attached to the filum terminale, and in 2.6% it was attached to the conus medullaris. In 25.7% of cases, no origin site was mentioned. In one case reported by Sundgren et al. [29], the tumor was extradural and recurred on three occasions after initial STR. In one case reported by Vural et al. [35], the tumor was a dumb-bell shape extending through the L4–L5 right intervertebral foramen that was lying medially to the psoas major muscle at the level of the corpus of the L5 vertebra.

Tumor recurrence after GTR without radiotherapy was reported in 4 of 184 cases (2.1%), with a mean interval of 35 months, either at the same or at a different site [7,16,18,32].

Tumor recurrence after GTR followed by radiotherapy was reported in one of eight cases (12.5%) after 14 years, which was associated with multiple cerebrospinal fluid metastases [33].

Tumor regrowth after STR without radiotherapy was observed in two of three cases (66.6%), with a mean interval of 150 months [11,22], whereas tumor regrowth after STR with radiotherapy was observed in three of 15 cases (20%), with a mean interval of 64.3 months (Table 2) [12,17,29].

Between the two biopsies [14,29] followed by adjuvant radiotherapy, the first case presented with tumor progression after 1 year with sacral and retroperitoneal extension, whereas the second case had a slight tumor increase at 14 months after radiotherapy. This patient underwent two

Table 2
Summary of recurrences/regrowth cases reported in the literature

Authors (Reference)	Initial localization	Initial RT (Y/N)	Recurrence/regrowth (y)	Further surgery	Second RT	Follow-up after last surgery
Recurrences in GTR cases without RT (4/184)						
Raftopoulos et al. [16]	L2–L3	No	Recurrence after 8 y	Reoperated (STR)	Yes	6 mo
Aggarwal et al. [18]	L4–L5	No	Recurrence after 2 y at a different site (L3)	NR	NR	NR
Singh et al. [32]	L2	No	Recurrence after 1 y			
Warrier et al. [7]	L3	No	Three recurrences at a different site (S1–S3) in 8 y	Reoperated three times (STR)	Yes	NR
Recurrences in GTR cases with RT (1/8)						
Thines et al. [33]	L4	Yes	Single metastasis in posterior cranial fossa after 7 y Recurrence (L4) and multiple metastasis after 14 y	Posterior fossa surgery	Yes	4 y
Regrowth in STR cases without RT (2/3)						
Böker et al. [11]	L3–L5	No	Regrowth after 12 y	Reoperated (STR)	No	6 mo
Strommer et al. [22]	L3	No	Regrowth after 13 y	Reoperated (GTR)	Yes	2 y
Regrowth in STR cases with RT (3/15)						
Taxy [12]	L2–L3	Yes	Regrowth after 9 y	Reoperated (GTR)	NR	NR
Batra et al. [17]	L2–L3	Yes	Regrowth after 1 mo			
Sundgren et al. [29]	L5–S1	Yes	Regrowth five times in 7 y	Reoperated three times (first GTR and second and third STR)	Yes	9 mo

RT, radiotherapy; GTR, gross total resection; STR, subtotal resection; NR, not reported.

separate therapeutic sessions with I-131 metaiodobenzylguanidine with no tumor progression over a 5-year follow-up.

On these grounds, radiotherapy may be considered only for incompletely resected tumors, even if resistance to this treatment has been documented [2].

In the literature, only 4 of 234 (1.7%) cases of metastasis from PGLs of the cauda equina have been reported [22,23,27,33].

In cases of recurrence or regrowth, no distinctive histopathological features of malignancy were described. In particular, Sundgren et al. [29] reported some nuclear pleomorphisms, atypia, and occasional mitoses; thus, neither histopathological characteristics nor radiological features may predict an unfavorable course.

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