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Case report

Sinonasal angiosarcoma

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ABSTRACT

Introduction: Angiosarcoma is a malignant tumour arising from endothelial cells that accounts for 1% of all sarcomas. The sinonasal site of angiosarcoma is exceptional.
Case report: The authors report a case of sinonasal angiosarcoma in a 53-year-old man. Despite wide resection by open surgery and postoperative chemoradiotherapy, several tumour recurrences were observed, requiring multiple operations.
Discussion: Only histological examination with immunohistochemistry is able to confirm the diagnosis (factor VIII, CD34 and CD31 antigens). Standard treatment consists of surgery with wide resection followed by radiotherapy. The authors discuss the treatment modalities and prognosis of this tumour.

1. Introduction

Sinonasal angiosarcoma accounts for 1% of all sarcomas [1]. The diagnosis of angiosarcoma can be suggested clinically in the presence of a bleeding tumour with contrast enhancement on computed tomography. However, the definitive diagnosis is based on histology, systematically associated with immunohistochemistry [2].

2. Case report

A 53-year-old diabetic and hypertensive man consulted for minimal unilateral right epistaxis with mucopurulent nasal discharge and partially blocked nose. Endoscopy demonstrated a polypoid mass of the right middle meatus with no palpable cervical lymph nodes. Complete resection of the mass was performed by endonasal surgery. Histological examination concluded on capillary haemangioma with no signs of malignancy. Three and a half months later, the patient was reoperated by endonasal surgery for tumour recurrence in the right middle turbinate (Fig. 1) with right middle meatal antrostomy and anterior ethmoidectomy. Histological examination comprising immunohistochemical staining was in favour of grade II angiosarcoma. The patient was treated by postoperative concomitant chemoradiotherapy: 50 Gy of radiotherapy and 6 cycles of cisplatin. Nine months later, the patient presented another recurrence in the medial wall of the orbit and was again reoperated via an endonasal approach. Surgery consisted of radical ethmoidectomy and was followed by 3 cycles of cisplatin and epirubicin. Five months after, a third recurrence in the left and right nasal cavities was resected via a lateral rhinotomy (Fig. 2), followed by chemotherapy (9 cycles of paclitaxel). Ten months later, the patient presented another naso-ethmoido-maxillary recurrence with no intracranial or orbital invasion and was reoperated via a lateral rhinotomy. As this extensive resection exposed the meninges of the anterior cranial fossa, the defect was filled by a frontalis myofascial flap and the skin defect was closed by a forehead flap. Gamma knife radiosurgery was then performed: 20 Gy to small residues on the medial walls of the orbit and ethmoid sinus (Fig. 3). Six months later, the patient presented with massive recurrence with externalisation of the tumour via the nostrils and surgical wound with frontal intracranial extension on computed tomography. In view of the cerebral extension of the tumour and the marked deterioration of the patient's general state, the patient was considered to be inoperable and died 10 days later.

3. Discussion

Angiosarcoma is a very rare malignant tumour accounting for almost 1% of all sarcomas [1]. The sinonasal site is exceptional, as only 24 cases have been reported since 1974 [2,3] in patients ranging in age from 8 to 81 years (our patient was 53 years old) with a male predominance (16 men for 8 women in the 24 reported cases). No known aetiology or risk factors have been identified, but telangiectasia, trauma and chronic oedema have been proposed [4,5]. Clinically, the tumour presents with similar symptoms to those of malignant sinonasal tumours: initially unilateral rhinological syndrome (epistaxis, blocked nose and rhinorrhoea) and, at a more advanced stage, tumour syndrome with invasion

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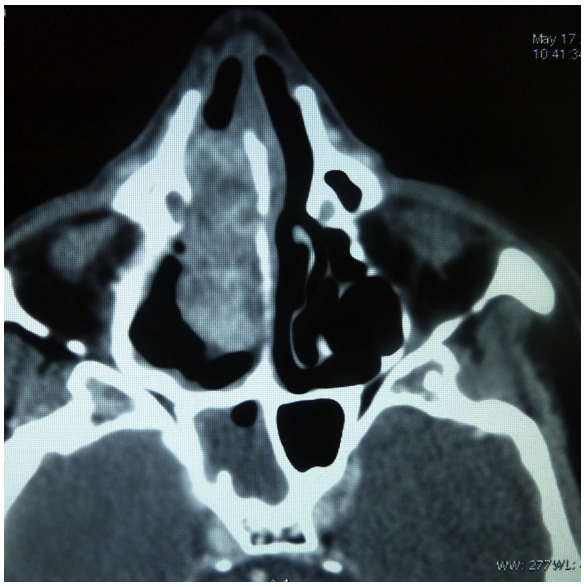

Fig. 1. Heterogeneously enhanced naso-ethmoidal tumour.

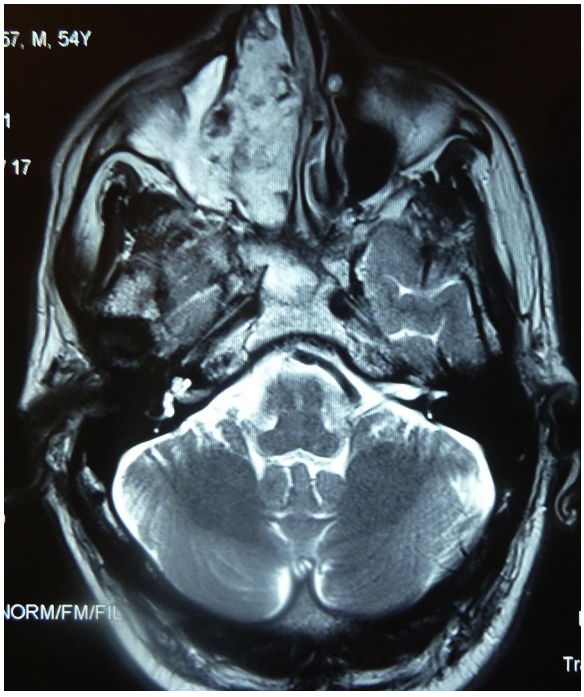

Fig. 2. Naso-ethmoid recurrence with heterogeneous high-intensity signal on the T2-weighted sequence.

of vital structures (eyeball, brain and infratemporal fossa). Epistaxis was the most common presenting complaint in the published cases and in the case reported here. On imaging assessment, the tumour presents variable dimensions (ranging from 2 to 8 cm in the published cases) with heterogeneous contrast enhancement and signs of bone destruction on CT scan. On MRI, the tumour presents a low-intensity signal on T1-weighted sequences and a heterogeneous high-intensity signal on T2-weighted sequences with intense, heterogeneous contrast enhancement. Histology with immunohistochemical staining is the only way to confirm the diagnosis. Macroscopically, the tumour is polypoid, nodular, violaceous red with a friable appearance and zones of haemorrhagic necrosis. Histological examination reveals a tumour composed of branched and anastomosed vascular structures lined by atypical endothelial cells (classified in grades) with a fibrous stroma containing necrosis and haemosiderin deposits [6]. The case reported here was classified as FNCLCC grade II (French cancer centre classification). Immunohistochemistry confirmed the diagnosis by showing positive staining for factor VIII, CD31 and CD34.

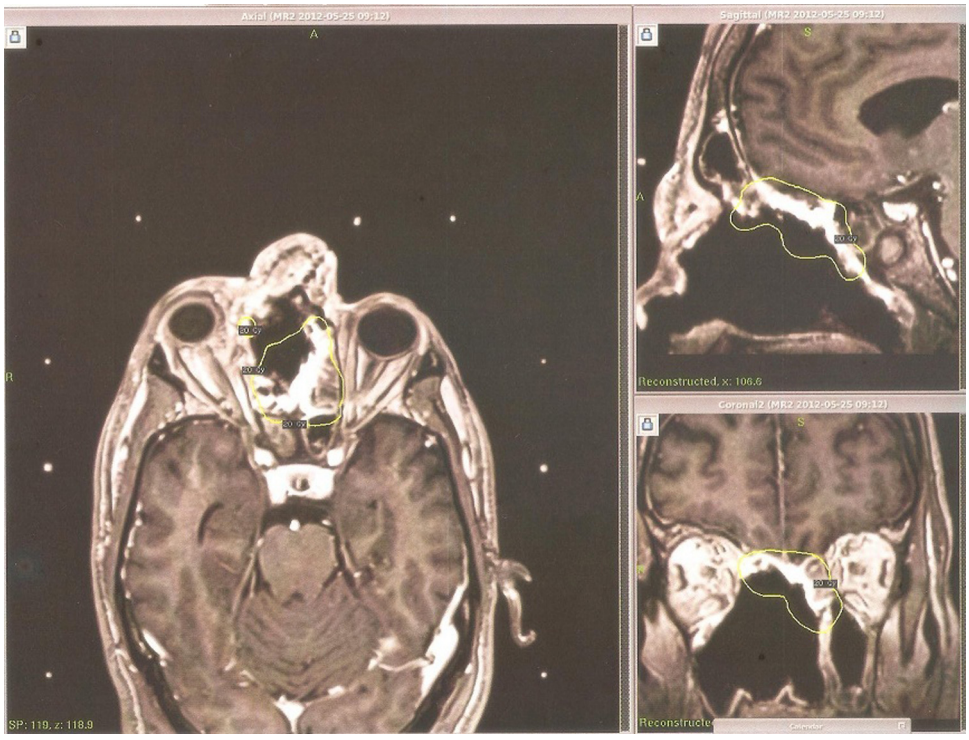

Fig. 3. Postoperative Gamma knife target volume.

The differential diagnosis includes haemangioma, intravascular papillary endothelial hyperplasia (Masson's disease), juvenile nasopharyngeal angiofibroma, haemangiopericytoma, Kaposi's sarcoma and malignant melanoma [6].

Treatment is based on surgery (adapted to the tumour site and extension) followed by radiotherapy. Other treatment modalities have been tried, such as chemotherapy, radiosurgery (gamma knife) [4], and interleukins [2–5]. However, no standardized therapeutic management has been defined in view of the limited number of published cases and the absence of comparative studies. The five-year survival rate is 22% (versus 12% for soft tissue angiosarcoma of the head and neck) and depends on the grade of differentiation of the tumour and the stage at diagnosis [7]. Distant metastases may occur during the first 24 months in 30% of cases; metastases involve bone, lungs, liver, skin or uterine cervix [3]. In our patient, the interval between diagnosis and death was almost 2 and a half years. The most commonly reported prognostic factors are: early diagnosis, grade of differentiation, the patient's age (over the age of 50), tumour size and resectability [2,8,9].

4. Conclusion

Sinonasal angiosarcoma is a rare tumour. It does not present any specific clinical features apart from its highly locally invasive and rapidly progressive nature. Only histological examination with immunohistochemistry can provide the definitive diagnosis.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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