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Case Report

Primary osteosarcoma of the sphenoid wing in a middle-aged woman with extensive intracranial extension: A case report

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الملخص

الساركومة العظمية الأولية التي تصيب قاعدة الجمجمة في منتصف العمر نادرة. نصف هنا حالة امرأة آسيوية تبلغ من العمر ٥٩ عاما تعاني مصابة بالخمول، ورعاف، وألم في الفك العلوي والسفلي الأيسر، وصداع. أظهرت فحوصات التصوير المقطعي المحوسب والتصوير بالرنين المغناطيسي وجود ورم كبير في الجناح الأيسر الكبير للعظم الوتدي مع ارتشاح محلي واسع النطاق وتمدد داخل الجمجمة. تم تشخيص الورم بأنه ساركومة عظمية عن طريق الفحص النسيجي. في هذا التقرير، نناقش الأعراض السريرية، والسمات الإشعاعية، والتصوير التشخيصي التفريقي لهذه الحالة.

الكلمات المفتاحية: الساركومة العظمية؛ قاعدة الجمجمة؛ العظم الوتدي؛ ورم الجناح الوتدي الأكبر؛ ألم الفك السفلي

Abstract

Primary osteosarcomas involving the base of the skull in middle-aged patients are rare. We describe the case of a 59-year-old Asian woman presenting with lethargy, epistaxis, left maxillary and mandibular pain, and headache. Computed tomography (CT) and magnetic resonance imaging (MRI) showed a large left greater sphenoid wing tumour with extensive local infiltration and intracranial extension. The tumour was diagnosed as osteosarcoma based on histological examination. In this report, we discuss the clinical presentations, radiological features, and imaging differential diagnoses of this case.

Keywords: Base of skull; Greater sphenoid wing tumour; Mandibular pain; Osteosarcoma; Sphenoid bone

Introduction

Osteosarcomas are malignant bone-forming tumours that mainly affect adolescents and young adults. They can be classified into primary and secondary osteosarcomas, with secondary osteosarcomas contributing to the majority of osteosarcomas diagnosed in the elderly.[1] Primary osteosarcomas in the older population, especially in the skull base, are rare.[2,3] When present in this location, the most common radiological differential diagnosis is atypical meningioma or dura-based metastasis. We report a case of primary osteosarcoma in the left greater wing of sphenoid bone with extensive intracranial extension and local invasion in a middle-aged woman.

Case report

A 59-year-old Asian woman presented with a month-long history of lethargy, left maxillary and mandibular pain, left-

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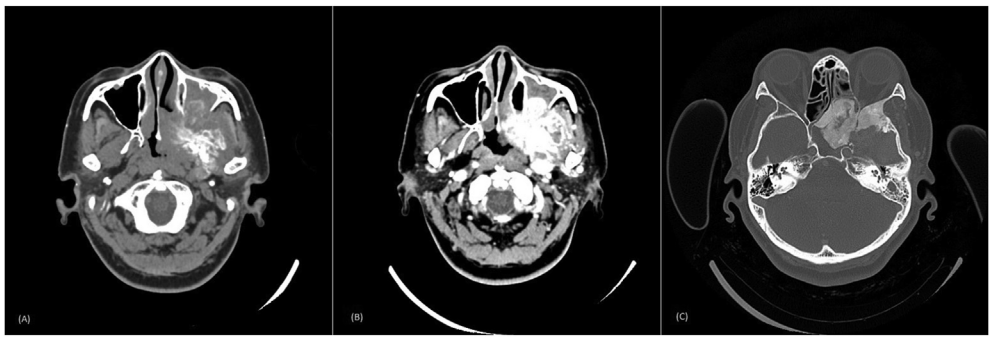

Figure 1: (A, B) Pre and post-contrast head CT images showing an ill-defined mass arising from the left greater sphenoid wing region which is avidly enhancing post-contrast. (C) Bone window showing hyperostosis of the left greater and lesser sphenoid wings and walls of the adjacent sphenoid and ethmoid sinuses.

sided headache, and recurrent epistaxis. The patient had underlying type 2 diabetes mellitus and hypertension, both of which were well controlled with oral medications. On admission, the patient appeared confused. Her neurological examination showed impaired memory and attention with a Mini-Mental State Exam score of 16/30. The patient exhibited right upper motor neuron facial weakness, weakness in the right upper limb with hyperreflexia, and right pronator drift. Her chest radiograph was clear, and no breast mass or neck mass was palpable. An attempted nasal scope's insertion was unsuccessful because the patient was restless.

Blood investigations showed a marked increase in alkaline phosphatase (ALP) levels with normal serum calcium levels. Head computed tomography (CT) demonstrated an ill-defined mass in the left greater sphenoid wing region which was avidly and heterogeneously enhanced on post-contrast images with an extension into the left middle cranial fossa. Coarse calcifications and areas of tumour necrosis were observed within the mass. Hyperostosis of the left greater and lesser sphenoid wings, as well as the walls of the adjacent sphenoid and ethmoid sinuses (Figure 1), was noted.

Magnetic resonance imaging (MRI) of the brain showed an isointense T1 and isointense T2 signal intensity mass arising from the left greater sphenoid wing which was heterogeneously enhanced in the post-gadolinium T1 sequence. Extension occurred in the left maxillary sinus, left middle and inferior nasal turbinates, posterior nasal fossa, and left middle cranial fossa. It indented onto the left cavernous sinus with encasement of the intra-canicular part of the left optic nerve and the petrous part of the left internal cerebral artery. In the left middle cranial fossa extension, there was a mass effect on the left temporal lobe, left basal ganglia, left thalamus, midbrain, and pons (Figures 2 and 3). The left lateral and third ventricles were compressed with a midline shift to the right (Figure 4). There was significant perilesional oedema involving the adjacent brain parenchyma on T2-weighted and fluid-attenuated inversion recovery sequences. There was no restricted diffusion on diffusion-weighted imaging (DWI) and apparent diffusion coefficient sequences, and it showed a low DWI signal. Extensive blooming artifacts in the susceptibility-weighted imaging (SWI) sequence were consistent with previous intratumoral haemorrhages and calcifications. Time-of-flight magnetic resonance angiography (MRA) showed superior displacement of the left middle cerebral artery and medial displacement of the left anterior cerebral artery (Figure 5).

The patient was referred to our neurosurgical colleagues for further workup and management. The patient's consciousness level deteriorated in the ward. Repeated head CT showed a worsening mass effect with left uncal herniation, cerebral oedema, and obstructive hydrocephalus, and the

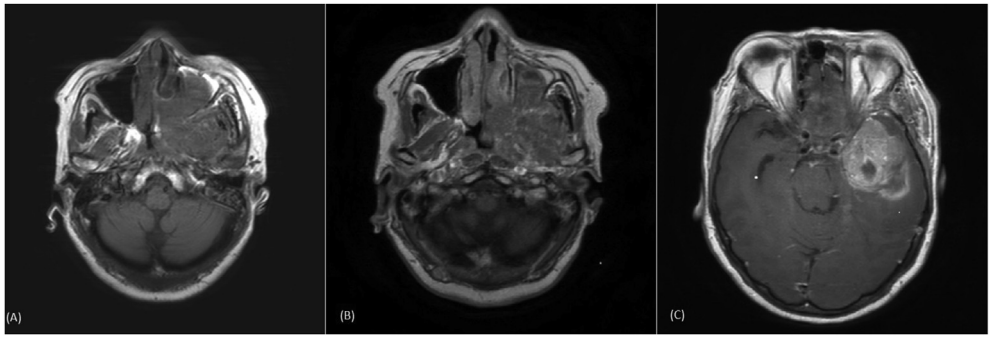

Figure 2: (A, B) Pre and post-contrast T1-weighted images showing heterogeneous enhancement of the lesion. (C) Post-contrast T1-weighted image showing invasion into the left middle cranial fossa causing mass effect onto the left temporal lobe. There is also an extension into the left carotid sinus with encasement of the intracanalicular part of the left optic nerve and the petrous part of the left internal carotid artery. Low signal intensity foci within the tumor in keeping with cystic changes.

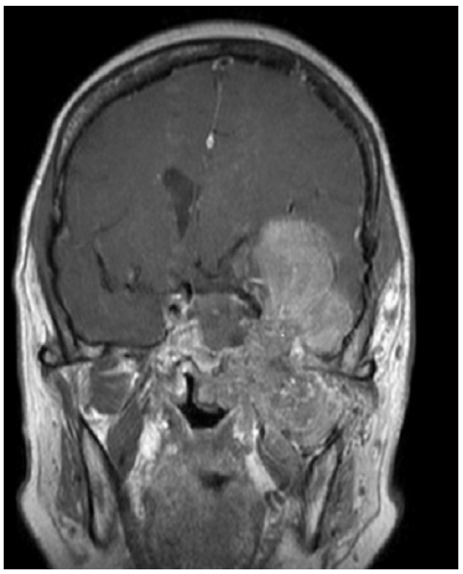

Figure 3: Post-contrast T1-weighted coronal image showing the extent of tumor infiltration from its epicenter in the left greater sphenoid wing into the surrounding structures and intra-cranially.

patient underwent an emergency craniectomy for decompression and tumour debulking. However, tumour debulking was abandoned because of the highly vascular nature of the mass. Immediate post-surgery head CT showed new intratumoral haemorrhages with worsening cerebral oedema and left uncal herniation.

Biopsy samples were successfully obtained intraoperatively. Histopathological examination showed a lace-like pattern of osteoid formation surrounded by malignant cells. Osteoid formation comprised irregular bony trabeculae and basophilic thin trabeculae. Malignant cells comprised atypical spindle cells with hyperchromatic nuclei. Epithelioid-appearing tumour cells and several multinucleated giant cells were also observed. These cells were stained positive for SATB2 and vimentin and negative for EMA, CKAE1/AE3, and GFAP. Overall, the histopathological findings were consistent with osteosarcoma, and a final diagnosis was made.

The patient was started on radiation therapy. She had several readmissions to control her worsening recurrent epistaxis from intranasal tumour extension. On follow-up, options for re-craniectomy with tumour debulking and radiotherapy were discussed. However, all the offered treatment options were palliative because of the high-grade nature of the malignancy. The patient opted for palliative care and was discharged on hospice care.

Discussion

We report a rare case of primary osteosarcoma of the greater sphenoid wing in a 59-year-old woman.

Based on her clinical presentation, radiological findings, and location of the tumour, we determined the most likely diagnosis as atypical meningioma of left greater wing of sphenoid. Another possible differential diagnosis was dura-based metastasis from a distant primary malignancy, which is common in her age group. Dura-based metastasis has a variable radiological appearance, presentation, and disease progression. The most common primary malignancies that cause dura-based metastasis are breast or lung malignancies. However, no evidence of primary malignancy was identified in her workup. The least likely differential was left sphenoid wing osteosarcoma.

Patients with osteosarcoma of the greater wing of the sphenoid usually present with facial mass, temporomandibular joint pain, ocular symptoms such as proptosis or decreased visual acuity, and headache.[4] Serum ALP is elevated, especially in the advanced stages of the disease. Our patient presented with left facial pain and headache, but without ocular symptoms, as the left optic tract was relatively preserved. Her confusion, impaired cognitive function, and unilateral weakness were due to the intracranial component exerting a mass effect on the adjacent brain parenchyma. Her recurrent epistaxis was due to bleeding from the intra-nasal extension of the tumour. Headaches, paresis, and change in mental status are common presenting symptoms of sphenoid wing meningiomas, epistaxis, and elevated serum ALP levels are rarely encountered. These clinical findings, the significant osteoid formation within the sphenoid bone on CT, and the aggressive nature of this tumour led us to include osteosarcoma in the differential diagnosis, even though it is rare.

Literature regarding the MRI features of skull base osteosarcoma is scarce. In a case series of 12 patients with a histopathologically confirmed cranium and skull base osteosarcoma by Luo et al., the predominant MRI features of skull base osteosarcoma were low to heterogeneous signal intensities on both T1- and T2-weighted images, heterogeneous or peripheral enhancement in post-gadolinium T1 sequence, and a low DWI signal.[5] In almost all of these cases, a dural tail sign was observed. Hayashi et al. reported a case of primary osteosarcoma of the sphenoid bone with extensive periosteal extension with MRI features of a ring-enhancing mass in the right sphenoid bone.[6]

In our patient, the tumour showed a heterogeneous appearance with isointense T1 and T2 signal intensities as compared to the grey matter, heterogeneously enhancing in post-gadolinium T1 sequence, and demonstrates a low DWI signal. No dural tail signs were observed.

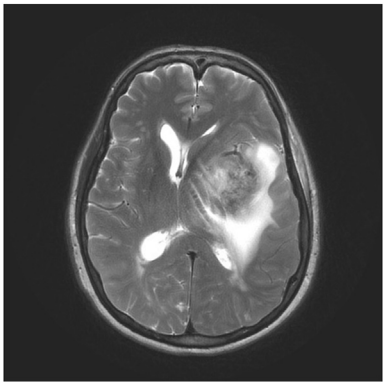

Figure 4: T2-weighted image showing intra-cranial tumor extension causing compression of the left lateral ventricle and midline shift to the right. Surrounding white matter edema.

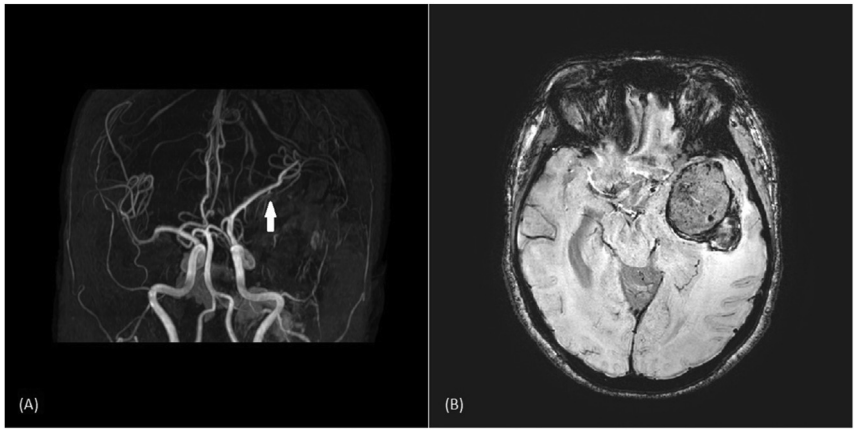

Figure 5: (A) Magnetic resonance angiography (MRA) sequence showing displacement of the left middle cerebral artery superiorly (Arrow). (B) Susceptibility weighted image (SWI) sequence showing blooming artifacts within and surrounding the lesion in keeping with calcifications and previous intra-tumoral hemorrhage.

Many of our MRI features overlapped with those of atypical meningiomas. Atypical meningiomas may arise from the skull base, causing hyperostosis of the adjacent bone; they may contain areas of tumour necrosis and show calcifications.[7] They are iso- to hypointense on T1-and T2-weighted sequences, heterogeneously enhancing in post-gadolinium T1-weighted sequences, and they typically demonstrate a dural tail sign.[8] Atypical meningiomas involving the nasal cavity have also been reported.[9]

Osteosarcomas are malignant bone-forming tumours that can be classified as primary or secondary osteosarcomas. Primary osteosarcomas can be further divided based on histologic subtypes, location, and anatomic relationship to the bone. The most common subtypes of primary osteosarcomas are osteoblastic, chondroblastic, and fibroblastic osteosarcomas.[10] Secondary osteosarcomas are osteosarcomas that develop from a previous underlying disease or condition, such as previous exposure to chemotherapy or radiation therapy, malignant degeneration of an underlying Paget's disease or fibrous dysplasia, bone infarct, giant cell tumour, or osteogenesis imperfecta.[11] The most common anatomical site for osteosarcoma formation is the metaphysis of the long bones; it is less commonly formed in the mandible, maxilla, and vertebra. Huvos et al. showed that 1.6% of all osteosarcomas arise from the skull, and approximately 13% of cases occurred in patients over the age of 40^2. In the Western population, this second peak of prevalence is mainly attributed to secondary osteosarcomas.[12]

Primary osteosarcomas in the elderly show an increased incidence of tumour formation in the axial skeleton compared to the younger population.[1,12] In a study conducted by Dae et al. involving 39 high-grade primary osteosarcomas in Asian patients above the age of 40 (median age of 53.1 years), the most common location was the femur (48.7%) followed by proximal tibia (20.5%) and pelvis (17.9%). No skull involvement has been reported.[13] Another multicentre study by Joo et al. involving 232 Asian patients above 40 years of age (median age of 50 years) showed that the femur was the most common site (41.1%), and only 4 patients showed skull involvement. Secondary osteosarcoma arising from previous radiation therapy was observed in 7 patients.[3] No Paget-associated secondary osteosarcoma was found in either study. This is attributed to the relative rarity of Paget's disease in the Asian population.[3,13]

The prognosis of primary and secondary osteosarcomas in older patients remains poor, with 5-year survival rates of 38.5% and 14.6%, respectively. Surgery in combination with chemotherapy is recommended for the treatment of secondary osteosarcomas, while surgery alone is recommended for the treatment of primary osteosarcomas. The effects of radiotherapy on the survival of older patients have not been well researched.[14]

Conclusion

We believe our case to be a de novo primary osteosarcoma, as our patient did not have any medical history or risk factors that predisposed her to develop secondary osteosarcoma. This osteosarcoma of the greater wing of sphenoid is a rare disease entity that shares MRI characteristics with atypical meningioma. The aggressive nature of this tumour, the presence of epistaxis, and a markedly elevated serum ALP level are the factors that made us consider this disease in our differential diagnosis. Histopathological confirmation remains the mainstay of diagnosis, and the prognosis of this disease remains poor.

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Conflict of interest

The authors have no conflict of interest to declare.

Ethical approval

The authors confirm that this study had been prepared in accordance with COPE roles and regulations. Given the nature of the study, the IRB review was not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Authors' contributions

Radiological findings were reported by ATM. The case report write-up was prepared by OWF and supervised by NKAK. OLW managed this case in the neurosurgical department. All authors have critically reviewed and approved the final draft and are responsible for the content and similarity index of the manuscript.

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