CASE REPORT

Primary Osteosarcoma of the Skull Base in a Pregnant Patient

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ABSTRACT

An 18-year-old female who was 14 weeks pregnant first noted vision loss in her left eye six days prior to admission. Ophthalmologic examination revealed 20/20 vision in the right eye and count fingers vision in the left eye. A marked relative afferent pupillary defect was present in the left eye. Ophthalmoscopic examination revealed a trace optic nerve pallor temporally in the left eye without associated disc oedema or haemorrhage. Magnetic resonance imaging of the brain demonstrated a heterogeneous mass of the left sphenoid sinus extending superiorly causing compression of the intracranial portion of the left optic nerve, and laterally into the left cavernous sinus. The patient underwent transphenoidal resection of the tumour whose histologic morphology revealed a grade 2 osteosarcoma. Following resection, vision returned to 20/20 in the left eye. The patient has been treated with chemotherapy with close monitoring of her pregnancy.

Keywords: compressive optic neuropathy, osteosarcoma, skull base tumour

INTRODUCTION

Osteosarcoma is the most common primary malignancy of bone. Although osteosarcoma can occur anywhere in the body, it most commonly affects the long bones. Histologically, it is characterised by osteoid or immature bone production by neoplastic spindle cells.^{1–3} Osteosarcomas of the head and neck are uncommon and comprise 6–10% of total cases.¹ They classically present in the third to fourth decades of life and occur as secondary tumours following radiation therapy or chemotherapy for another tumour. They most frequently involve the mandible and less commonly the sphenoid sinus and maxilla.^{1–4} We report a pregnant patient with a primary skull base osteosarcoma who presented with rapid unilateral vision loss.

CASE REPORT

An 18-year-old female who was 14 weeks pregnant first noted vision loss in her left eye six days prior

to admission. She denied eye pain, pain with eye movements, parasthesias, weakness, recent fevers, or weight loss; however she reported a dull, intermittent, left-sided headache for four days. Her past medical history is significant only for an uncomplicated pregnancy course.

Ophthalmologic examination revealed 20/20 vision in the right eye and count fingers vision in the left eye. A marked relative afferent pupillary defect was present in the left eye. Motility was full. Slit lamp examination was unremarkable. Ophthalmoscopic examination revealed trace optic nerve pallor temporally in the left eye without associated disc oedema or haemorrhage.

Magnetic resonance imaging of the brain demonstrated a heterogeneous mass of the left sphenoid sinus extending superiorly, causing compression of the left optic nerve, and laterally into the left cavernous sinus (Figure 1). The patient underwent trans-sphenoidal resection of the tumour. Soft tissue biopsies were sent for frozen and permanent sections and revealed a grade 2 osteosarcoma. Specifically, the specimen demonstrated a cytologically malignant

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FIGURE 1 (A) Sagittal T1-weighted image demonstrates a heterogeneous, intermediate-to-low signal intensity mass centred in the sphenoid sinus (arrow) abutting the clivus. (B) Axial T1-weighted image at the superior aspect of the lesion shows the mass abutting the intracranial and intracanalicular portions of the left optic nerve (arrow).

neoplasm with spindle and ovoid cells arranged in intersecting fascicles. Reactive woven bone and extracellular matrix suspicious for neoplastic bone were also present (Figures 2 and 3). Following resection, vision returned to 20/20 in the left eye within two days.

Technetium-99 whole-body bone scan demonstrated isolated uptake in the base of the left skull, however computed tomography (CT) of the chest was negative for metastases. Systemic chemotherapy was initiated with the exception of methotrexate given its contraindication in pregnancy. The patient has undergone two cycles of cisplatin and doxorubicin and one cycle of etoposide and ifosfamide with close monitoring of her pregnancy.

DISCUSSION

Osteosarcomas of the head and neck are rare and usually present in the third to fourth decade of life, almost a decade after the most common presentation of long-bone tumours.^{2,4,5} Most common sites of involvement include the mandible and less commonly the maxilla and sphenoid sinus. The present case is unique in that this pregnant 18-year-old patient presented with only progressive vision loss and was found to have a primary osteosarcoma of the skull base with compression of the left optic nerve. Given the size and location of the lesion, it is likely that the vision loss in the left eye occurred gradually over an extended period of time, but was simply noted acutely by the

FIGURE 2 Low-power magnification shows malignant spindle cells arranged in fascicles. High power magnification demonstrates increased mitotic activity (insert). *Note:* Figures 2 and 3 of this article is available in colour online at www.informahealthcare.com/oph.

patient six days prior to admission. Complete surgical resection is the primary method of treatment of the primary tumour,^{1-3,5} however the anatomy of the head and neck often makes complete resection difficult.⁶ Adjuvant chemotherapy typically consists of cisplatin, doxorubicin, and high-dose methotrexate.^{1,2,5} Treatment options were limited for our patient due to her gravid status, as methotrexate is contraindicated in pregnancy. Whether this will affect

FIGURE 3 (A) Interface of spindle cells with hypercellular areas with neoplastic bone. (B) High power magnification demonstrates neoplastic bone with osteoblastic rimming.

prognosis is unknown and long-term follow up is needed.

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