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Case Report

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OSTEOSARCOMA OF THE FRONTAL BONE: A CASE REPORT

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ABSTRACT

Osteosarcoma as a primary tumour of the skull is very rare. The tumour biology slightly differs from osteosarcoma occurring in the extremities, with a lower tendency to metastasize distantly. Treatment of choice is surgery where possible, then adjuvant chemoradiation because achieving negative surgical margins at the head and neck region is extremely difficult. There is high chance of local tumour relapse. We report a case of a 48-year old female who presented with a 15-month history of rapidly increasing frontal mass and progressive visual loss. Histology confirmed the tumour to be osteogenic sarcoma. She underwent tumour debulking followed by adjuvant palliative chemotherapy.

KEYWORDS: Osteosarcoma, tumour, osteogenic.

INTRODUCTION

An approximate incidence of 2% of all skull tumours is constituted by primary osteogenic sarcoma of the skull.[1] It is very rare, constituting less than 10% of all osteosarcomas and so far, less than 200 cases have been reported in the literature.[2,6] The incidence of osteogenic sarcoma of the skull is said to peak around 20-39 years of age.[7] Osteosarcomas of the skull are more frequently secondary malignancies that occur commonly around the base of the skull and calvaria following earlier treatment with either radiation therapy or chemotherapy for another malignancy.[1,8]

In this case report, we present a case of osteogenic sarcoma of the frontal bone in a 48-year old female, as well as describe the clinical/radiological features and treatment.

CASE PRESENTATION

A 48-year old housewife who presented to the neurosurgery outpatient clinic with a fifteen month history of progressive frontal swelling, progressive visual loss and recurrent nasal blockage. There was associated recurrent frontal headache but no vomiting, convulsion, irrational behavior, cough or weight loss. There was no antecedent history of trauma or exposure to ionizing radiation. On examination, she was fully conscious and well oriented, with a Glasgow coma scale (GCS) score of 15. There was a mass overlying the frontal aspect of the skull; measuring 8x10x8cm, hard and irregular but firmly adherent to the underlying structures, with ill-defined margins. There were no palpable cervical lymph nodes. Other systemic examinations showed normal findings.

A cranial computed tomography (CT) scan done revealed a hyperdense lesion involving the frontal region, with destruction of the frontal sinuses and extension into the anterior cranial fossa, ethmoidal, sphenoidal and maxillary sinuses. Other staging investigations done were essentially normal. She had a frontal craniectomy and tumour excision with bilateral ethmoidectomy. The intra operative findings were those of a fibro-osseous tumour involving both frontal and ethmoid sinuses on both sides, with thick secretions in both maxillary antra. The histology revealed osteogenic sarcoma. She was thereafter commenced on cyclical cytotoxic chemotherapy about 4 weeks post-op using intravenous (IV) Vincristine 1.5mg/m^2, IV Doxorubicin 50mg/m^2 and IV Cyclophosphamide 600mg/m^2. She is being planned for cranioplasty and adjuvant radiation therapy after completing the chemotherapy.

DISCUSSION

Osteogenic sarcoma is the commonest primary malignant tumour of the bone and commonly occurs in the long bones of the extremities.[9,11] Osteosarcomas arise from osteoid-producing mesenchymal cells. The natural history of osteosarcomas arising in the head and neck bones somewhat differs from that arising in other skeletal bones, having a lesser tendency to metastasize distantly and a better 5-year survival rate.[12] The presence of suture lines, foramina and thin cortices of the craniofacial bones makes local and intracranial spread of head and neck osteosarcomas easy, hence majority of the

patients present with very advanced diseases.[12] Symptoms vary from painless slowly growing frontal mass, mild discomfort, to excruciating headaches. Majority of skull osteosarcomas are painless unlike those of the extremities.[13,14] Other symptoms may include exophthalmos, cranial nerve deficits and loss of vision, hearing impairment, recurrent convulsions and headaches. Osteogenic sarcomas of the skull and facial bones rarely metastasize to distant sites, but more often recur locally.[15]

Histopathologically, osteosarcomas are pathognomonically characterized by excessive production of irregular and immature osteoid by spindle cells.[14] Pathologic subtypes of osteosarcoma include osteoblastic, chondroblastic, fibroblastic osteosarcomas, as well as rarer subtypes like periosteal, parosteal, telangiectatic and small cell osteosarcomas.[16] These have no remarkable impact on the prognosis of skull osteosarcomas.[17] Osteosarcomas can also be graded microscopically into high, intermediate or low grade.

Radiologically, osteosarcomas are commonly diagnosed using CT scan with bone window, where they show abnormal bone growth with lytic areas and periosteal remodeling.[18] Magnetic resonance imaging (MRI), especially with contrast, could be used to rule out soft tissue or intracranial involvement. Osteosarcomas commonly enhance homogenously on MRI with contrast, with well defined margins, usually isointense on T1-weighted and hypointense on T2-weighted.[19] Other radiological investigations like chest xray, chest CT scan, abdomino-pelvic ultrasound scan, could be used to rule out metastatic disease.

The mainstay of treatment for frontal bone osteosarcoma, just like osteosarcomas in the extremities, is surgical excision.[20] Surgical resection should aim for complete excision with negative margins, because this is associated with better prognosis.[14] Occasionally, aggressive surgery might lead to some functional or structural deficits due to the peculiar anatomy of the head and neck region. Therefore complete surgical resection is rarely achievable; hence adjuvant therapy is almost always necessary in the majority of cases.[21] Further surgical resections to achieve negative margins could be attempted in a few cases. Adjuvant radiation therapy using photons or electrons, to a dose between 30-60Gy, has been largely shown to prevent local tumour recurrence and improve survival in cases of subtotal excision.[22] Chemotherapy has also been shown to improve overall outcomes when used either neoadjuvantly (to decrease tumour bulk and make it operable) or adjuvantly following positive surgical margins.[23,24] Commonly used chemotherapy agents include Methotrexate, Doxorubicin, Ifosfamide, Cisplatin, Etoposide, Cyclophosphamide and Vincristine.[25,27]

CONCLUSION

Osteogenic sarcoma of the skull is rare and usually poses a great diagnostic and treatment challenge. Most cases are metastases to the skull bone but primary osteosarcoma should also be suspected based on the presentation. Total surgical excision followed with adjuvant chemoradiation is the currently recommended treatment.

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