CASE REPORT

GIANT CELL TUMOUR OF THE OCCIPITAL BONE IN A 13-YEAR OLD MALE

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Giant Cell Tumours (GCT) are usually found at the extremities of the long bones and their presence in the skull being less than 1%. In the skull, sphenoidal bone and temporal bone are the commonest sites. There have been very few reports of GCTs of the occipital bone. Total excision surgery is the ideal treatment of choice. If surgery poses a problem, then adjuvant radiotherapy can be administered too. We present a case of 13-year-old male child who was diagnosed with GCT of the occipital bone. He was successfully operated and is symptom free 6 months post his surgery till now.

Keywords: Giant Cell Tumour; Occipital bone; Osteoclastoma; Excision Surgery

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INTRODUCTION

Sir Astley Cooper in 1818 first described Giant Cell Tumour (GCT). Since then, it has been given many names such as Osteoclastoma, Osteoblastoma, tumour of myeloplaxus and myeloid sarcom.[1] Though the tumor shows benign histologic features it can be locally aggressive too with destructive borderline malignant potency.[2] GCTs develop through endochondral ossification and hence are commonly found at the extremities of the long bones like distal femur, proximal tibia and distal radius.

They have also been seen in the ankle region and foot. GCTs of the skull are unusual and contribute less than 1% of the cases overall.[1–4] Occurrence of GCTs in the occipital bone is extremely rare with only few cases being reported in the literature.[5–12] We present an interesting case of a 13year old male child who presented to us with complaints of headache and swelling in the head, was diagnosed as GCT of the occipital bone. He was successfully treated with radical surgery and has shown no signs of recurrence 6 months post his surgery.

CASE PRESENTATION

A 13year old male child presented to us with complaints of headache and swelling in the right occipital region for 9 months and multiple episodes of vomiting for 1 month. The headaches were paroxysmal and recurrent in nature with the swelling gradually increasing in size since the past 2 months. Examination revealed the swelling to be 5×5 cm in size, non-mobile, non-tender and firm in consistency. Right sided ataxium was seen too. Neurological examination discovered a positive past pointing test. He had no significant history of trauma, fever or discharge. Magnetic Resonance Imaging (MRI) revealed a well-defined large expansile lesion measuring 5.2×5.6×5.3 cm compressing the 4th ventricle, right cerebellum and quadrigeminal cisterns (Figure-1A + Figure-1B). Magnetic Resonance Angiography (MRA) did not reveal high vascularity while Magnetic Resonance Venography (MRV) showed no flow void in the right transverse, sigmoid and right internal jugular vein.

The patient underwent a retromastoid suboccipital craniotomy with a complete excision of the tumour in the form of en bloc removal. The tumour was normal in color and moderately vascular in nature. Pathological evaluation of the specimen revealed numerous osteoclasts like cells with surrounding mononuclear cells and highly vascular stroma with thin-walled vascular channels (Figure -2 +Figure -3). Immunochemical staining revealed positivity for CD 34 with presence of vimentin and Kp-1. A diagnosis of GCT of the occipital bone was made. The post operative period was uneventful with no added neurodeficit. At the 6 months follow up, the patient is doing fine with no signs of recurrence and is able to lead a normal life.

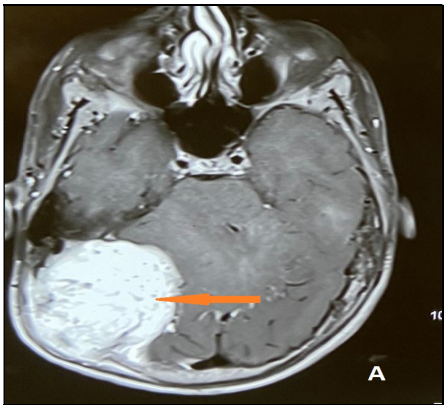

Figure-1A: MRI (axial) T2 flair weighted image shows a hyperintense well defined expansile lesion (red arrow) measuring 5.2×5.6×5.3 cm in the right occipital region

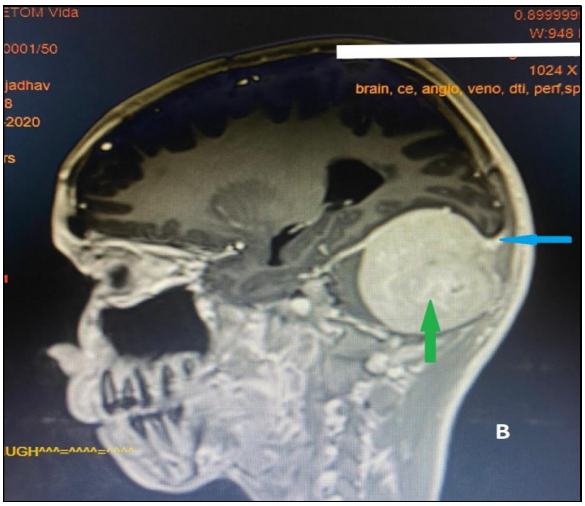

Figure-1B: Post Gadolinium enhanced MRI (sagittal) T1 weighted image shows lesion (green arrow) with dural tail (blue arrow).

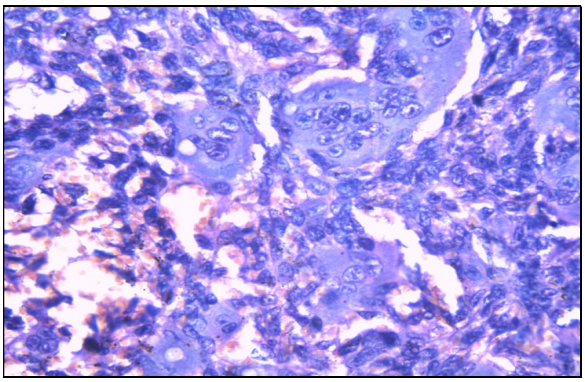

Figure-2: H& E staining of the tumour (magnification x20) shows osteoclastic giant cells with surrounding mononuclear giant cells and thin-walled vascular channels. Mitosis and necrosis not seen.

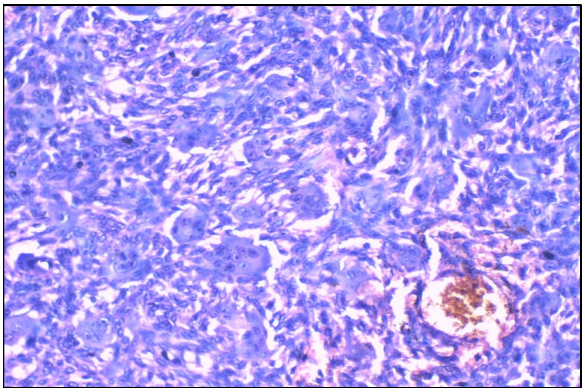

Figure-3: H& E staining of the tumour showing mononuclear cells having vesicular nuclei with prominent nucleoli. Stroma is highly vascular with the presence of many thin-walled blood vessels. Mitosis or necrosis is not seen.

DISCUSSION

Giant Cell Tumour in the skull is a rare phenomenon with sphenoidal and temporal bones being the two commonest sites. The process of development in these bones is through endochondral ossification which is akin to the process of development of GCTs and hence explains the occurrence. Occipital bone develops through intramembranous ossification. The presence of GCTs in membranous bones remains a mystery even though various mechanisms like occurrence of metaplasia in primitive connective tissue and presence of aberrant cells have been proposed.[2,6,7] A thorough review of the literature revealed very few reports of occipital bone GCTs.[4–11] Our patient too was diagnosed with GCT of the occipital bone. Giant Cell Tumours unusually show preponderance for the Chinese population and females with many of them occurring in the age groups of 20–50 years. Their incidence in skeletally immature patients (<14 years) is reported to be a measly 1–3%.[1] The previous reports too showed a female preponderance, but our patient was a male child aged 13 years.[4,5,8–12] There have been only 2 cases of males being reported and one of them being the youngest at 12 years old.[6,7]

The most common clinical presentation is an enlarging mass associated with pain and swelling of the affected region over a period of few weeks to years.[8] Our patient presented with headaches, vomiting and an enlarging mass over the occipital region with no neurodeficit. Since plain radiographs of the skull present GCTs as radiolucent lesions making it difficult to differentiate them from other radiolucent lesions, both Computed Tomography (CT) and MRI are used for radiological investigations. MRI provides a better characterization and delineation of the tumour than CT. CT alone is inadequate for the precise diagnosis and segregation of GCTs from other similar looking masses or tumours, such as giant cell reparative granulomas (GCRG) or osteitis fibrosa (brown tumour). These tumours might be vascular, and a prior knowledge of the vascularity is essential for the differential diagnosis along with helping the surgeons to plan and improve the interventional procedures.[4,13,15] In our patient, Magnetic Resonance Angiography (MRA) did not reveal high vascularity while Magnetic Resonance Venography (MRV) showed no flow void in the right transverse, sigmoid and right internal jugular vein. These findings were confirmed intraoperatively. None of the previous reports performed MRA/MRV.

The pathological differential diagnosis of GCTs is widespread and includes GCRG, osteoblastoma, Aneurysmal Bone Cyst (ABC) brown

tumour of hyperparathyroidism, chondroblastoma, nonossifying fibroma, foreign body reaction, and osteosarcoma with abundant giant cells. A true GCT contains an enormous number of giant cells distributed diffusely in a background of mononuclear cells which are predominantly round, oval, or polygonal. The stroma of most GCTs is vascular with abundant thin-walled capillaries and small areas of haemorrhage intermixed.[1] The histopathological evaluation of our patient was consistent with the above findings. Though immunohistochemistry isn't mandatory for the diagnosis it may reveal positive stain for MMP – 13, CD 68, P53, PDGFA, cyclin D1 and C-kit. Only 2 of the previous cases performed immunohistochemistry.[9,10,14] In our institute, we perform immunohistochemistry as a standard protocol for all specimens.

Complete surgical excision is the ideal treatment of choice for GCTs but location of the tumour, poses considerable threat to the patient due to the presence of critical neurovascular structures around. For unresectable/partially resectable tumours, adjuvant chemotherapy can be administered. But its role is still controversial and debatable with some reports suggesting that GCT isn't radiosensitive, and that radiation can trigger a sarcomatous transformation in the residual tumour tissue. Others recommend a single course of moderate dose super voltage radiation in achieving a high success rate and simultaneously reducing the probability of malignant transformation. A combination of surgery and radiotherapy has also been used.[2,8,9] Our patient underwent a total excision of the tumour, hence radiotherapy wasn't required and has shown no signs of recurrence till date.

CONCLUSION

GCTs of the skull are a rare entity. When they do occur, sphenoidal and temporal bones are the preferred sites due to similar developmental process. Presence of the tumour in the occipital is extremely rare and should be handled with caution. Total gross resection is the ideal treatment but poses a threat to the patient due to existence of critical structures around. If surgery isn't feasible or has been performed sub totally, radiotherapy can be administered.

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