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Case Note

An Unusual Presentation of Giant Cell tumour (osteoclastoma)

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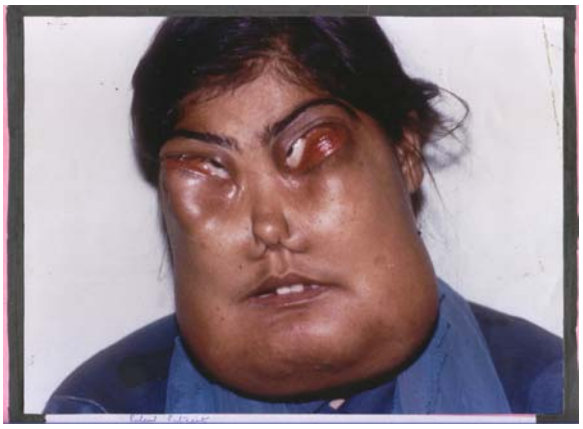
Abstract:
The number of well-documented true giant cell tumours arising in any of the craniofacial bones is small, but they do exist. A 19 year old female, Ms. KS, presented with complain of progressive enlargement of facial bones especially jaw bones, then orbit symmetrically since the age of 7. There was bilateral gross enlargement of mandible, maxilla, orbital walls, causing displacement of eye medially and upwards. The visual acuity of both eyes were 6/36 and 6/18 with best correction. Extra ocular movements were restricted because of bony growth and conjunctiva over inferior fornix were keratinized due to exposure. Fine needle aspiration (FNAC) from the side of bony growth showed plenty of osteoclasts with multinucleated giant cells. The level of serum alkaline phosphatase were highly increased. She underwent orbitotomy and a part of tissue was sent for biopsy which revealed multiples of mononucleargiant cells and tumour cells.

Key Words: osteoclastoma, craniofacial bones, visual acuity, orbitotomy.

The number of well-documented true giant cell tumours (osteoclastoma) arising in any of the craniofacial bones is small but they do exist. Several cases of giant cell tumours involving jaw bones, skull, petrous temporal bones and sphenoid etc were reported[1234]

Case Report
A 19 year old female, resident of Jhapa, Nepal attended Eye camp and subsequently referred to Ophthalmology, Tribhuvan University Teaching Hospital. She presented with the complaints of progressive enlargement of facial bones, especially jaw bones then orbit symmetrically since the age of 7. The growth became rapid since last 2 years.



The general condition of the patient was fair. There was bilateral, gross enlargement of mandible, maxilla, inferior and lateral orbital walls. The eyes wee displaced medially and upwards and nose was pinched with nasal intonation of voice of the patient. The growth was bony hard, non- tender and non-compressive. On neurological examination, higher mental function was normal and the cranial nerves were intact. There were no sensory or motor deficit. There was no cerebellar and meningial sign.

Similarly on ocular examination, visual acuity were 6/36 in both eyes and 6/18 with best correction. Extra ocular movements were restricted in lateral, up and down gazes. Conjunctiva in both eyes over the inferior fornix were exposed due to underlying growth and were keratinised. Fundus in both eyes were unremarkable and pupillary reflexes were normal.

Investigations
X-ray skull showed enlarged facial bones, including maxilla and mandible with mixed type of lesions, sclerosis and luscent areas. There was frontal bossing mainly due to pertuberant frontal sinuses. Teeth were floating but the bones of the base of the skull and skull vault were normal.

Fine needle aspiration cytology showed plenty of osteoclasts like giant cells with round to oval nucleus,

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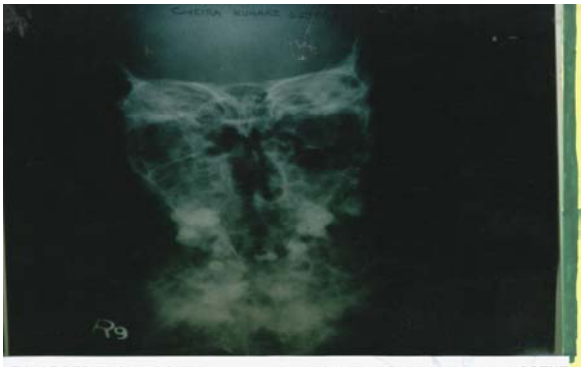
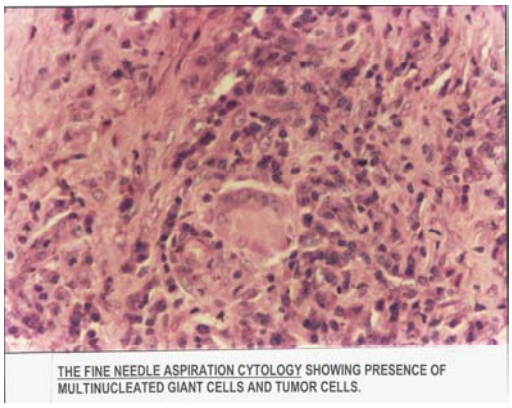


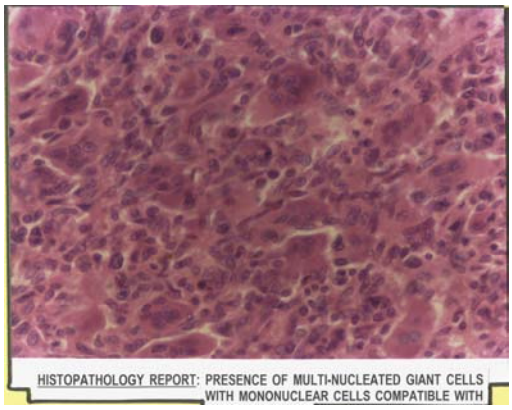
ENLARGED FACIAL BONES AND MANDIBLES WITH SCLEROTIC AND LUSCENT AREAS ALONG WITH FLOATING TEETH.

varying from 10-100 nuclei. Few stromal cohesive clusters of spindle cells are also present with intercellular collagen in haemorrhagic background.

The serum calcium level was normal but the level of serum alkaline phosphatase was highly increased (306u/l).



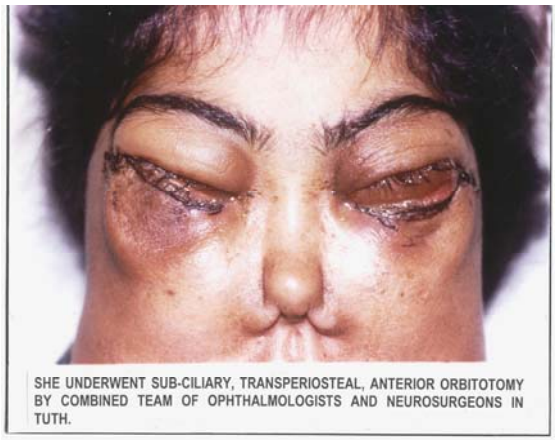
THE FINE NEEDLE ASPIRATION CYTOLOGY SHOWING PRESENCE OF MULTINUCLEATED GIANT CELLS AND TUMOR CELLS.



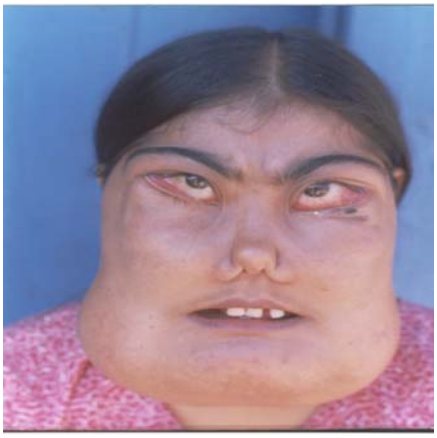
HISTOPATHOLOGY REPORT: PRESENCE OF MULTI-NUCLEATED GIANT CELLS WITH MONONUCLEAR CELLS COMPATIBLE WITH

Management

She underwent subciliary, trasperiosteal anterior orbitotomy by combined team of ophthalmologists and neurosurgeons. The biopsy of the tissue revealed multiples of multinucleated giant cells and tumour cells. Hence the final diagnosis was made as osteoclastoma of facial bones with the orbital involvement. After the excision of the bony orbital growth, there was no recurrence



SHE UNDERWENT SUB-CILIARY, TRANSPERIOSTEAL, ANTERIOR ORBITOTOMY BY COMBINED TEAM OF OPHTHALMOLOGISTS AND NEUROSURGEONS IN TUTH.



Discussion

Giant cell tumours are distinctive neoplasms because they are characterized by a profusion of multinucleated giant cells scattered throughout a stroma of mononuclear cells. The stromal cells are the neoplastic element, not the giant cells. Since the giant cells have some similarity to osteoclasts, these neoplasms have been inappropriately called 'Osteoclastoma'. The neoplastic element or the mononuclear cells are origined from mesenchymal cells and they are the progenitors of the giant cells[5].

The mononuclear phagocyte system (MPS) consists of cells in different systems of the body, as in the blood vascular system, it is called monocyte and macrophages in tissues of different parts of the body. Monocyte chemoattractant protein 1 (MCP-1) is a potent chemotactic factor specific for monocytes. In search of relevant cytokines that may enhance the recruitment of these reactive cells, a group of scientists evaluated the localization and regulation of MCP-1mRNA and protein in giant cell tumour (GCT) by using northern blot analysis, in situ hybridisation and immunohistochemistry. They also

determined whether conditioned medium obtained from GCT cultures can recruit human peripheral monocytes (CD68+) in an in vitro chemotactic assay. Using northern blot analysis, they detected the specific gene transcript for MCP-1 in all GCT samples tested. In situ hybridisation and immunohistochemistry revealed that both MCP-1 gene transcript and protein were consistently present in the cytoplasm of stromal-like tumour cells of GCT[6].

In view of the management of the disease, wide excision of the enlarged tissue along with reconstructive surgery is considered as well as life lone follow-up is also equally important[7].

Conclusion

Giant cell tumour (Osteoclastoma) is considered as true neoplasm arising from undifferentiated cells of bone marrow. Such tumour appear in children usually under 5 years of age and occur most commonly at the ends of long bones but are occasionally seen in the skull particularly in the jaws. A few were reported in sphenoid[8,9,10] frontal bone, ethmoid labyrinth and temporal bone.

Acknowledgement

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