

Chordoma in the sella turcica

A Khaled¹, A Joarder², M Chandy³, T A Nasir⁴

Introduction: A chordoma is a slow-growing malignant tumour with a metastatic potential developing from notochord remnants. The incidence is about 1 % of all malignant bone tumours with a male to female ration of 1 : 1. Most patients are in their fifth to seventh decades but patients presenting with skull or vertebral tumours are younger. Chordomas have a protracted course and symptoms are usually present for more than 1 year. The symptoms related to the size and location of the tumour are varied: headache, increased intracranial pressure or cranial nerve involvement for sphenoccipital tumor, pain and neurologic symptoms for vertebral tumours and pain, mass, neurologic symptoms or rectal and bladder dysfunction for sacral and sacrococcygeal tumours.

Intracranial chordomas have been described associated with Maffucci's syndrome, hemangiopericytomas, neurofibromatosis or tuberous sclerosis or presenting as a second primary malignant lesion².

Histogenesis : The notochord forms the first axial skeleton during embryogenesis a rod of ectodermal cells is presumed to act as an embryonic organizer for the chondrification and segmentation of the mesenchymal elements of the vertebral bodies. During the fifth week, this becomes enclosed within the developing vertebral column and is divided in segments.

The notochord regresses during fetal life but there are variations in its persistence and regression. Remnants are left in the nucleus pulposus, particularly in the upper and lower ends of the column and also in the midline of the sphenoccipital region².

Chordomas arise along an archipelago of this notochordal remnants extending from their rostral reaches in the region of the sella turcica to their caudal extreme in the sacrum¹.

Case report

A 44 years old nondiabetic, normotensive, Bangladeshi female was admitted through ER with the complain of sudden severe headache, vomiting for 1 day and loss of vision for 1 week. She also complained gradual loss of vision, mild headache and menstrual disturbance for the last 6 months. On examination her vitals were stable and glassgow coma scale was 15/15. Examination of eye revealed both pupils were 2 mm and equally responsive to light, normal anterior segment of both eye, visual acuity of right eye 6/24 and left eye 1/60, perimetry of right eye shows loss of temporal field along with relative upper and lower quadrant field defect, perimetry of left eye was not possible due to poor vision. Fundic examination of left eye shows pallor of the disc and right eye shows mild pallor of the disc. Intraocular pressure of right eye was 21 mm Hg

and left eye 18 mm Hg. MRI examination of brain showed well delineated approx 4.5x3.7x3.5 cm sized fairly large nodular infiltrating space occupying lesion in the suprasellar cistern extending into intrasellar and parasellar region causing significant mass effect over the surrounding structures. Transsphenoidal tumour decompression was done and tissue was sent for histopathology. Grossly, the tumour tissue was gelatinous and grayish white totally measuring around 1 cc. Microscopically, the tumour tissue show proliferation of large cells with abundant pale vacuolated cytoplasm and uniform nuclei arranged in sheets and cords in a background of myxoid stroma. Few physaliphorous cells and areas of calcification are also

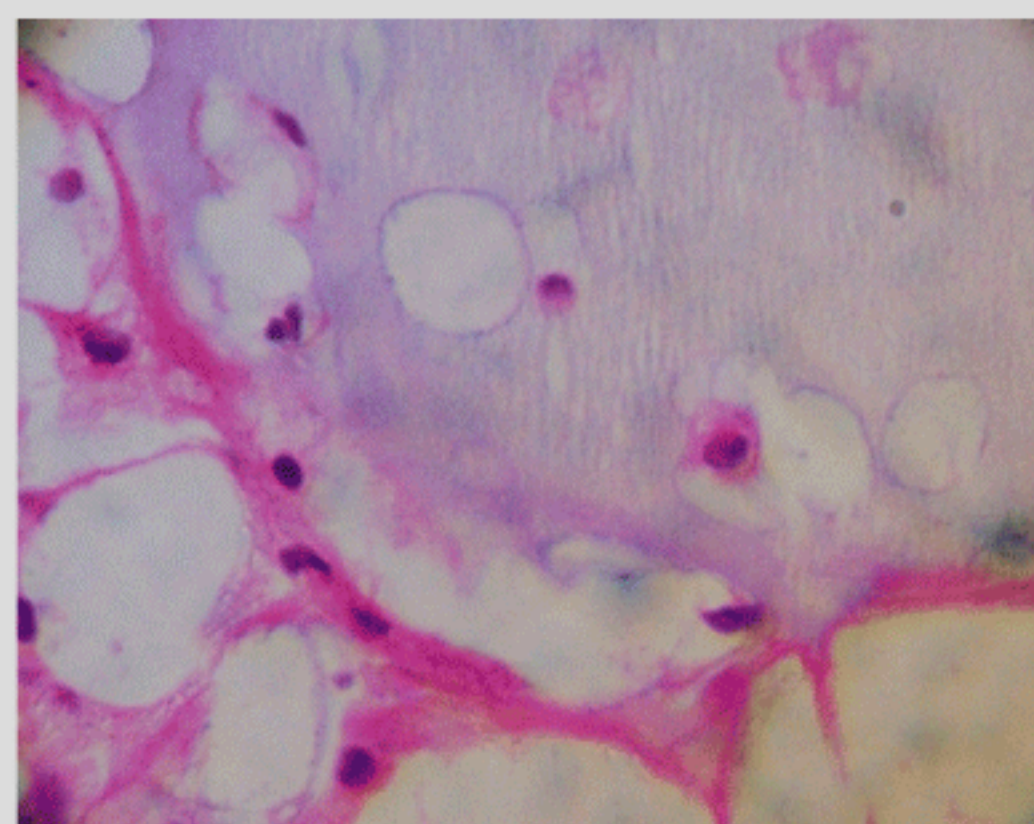


Figure 1: Chordoma. Large vacuolated cells and physaliphorous cells in a myxoid stroma (HE x40).

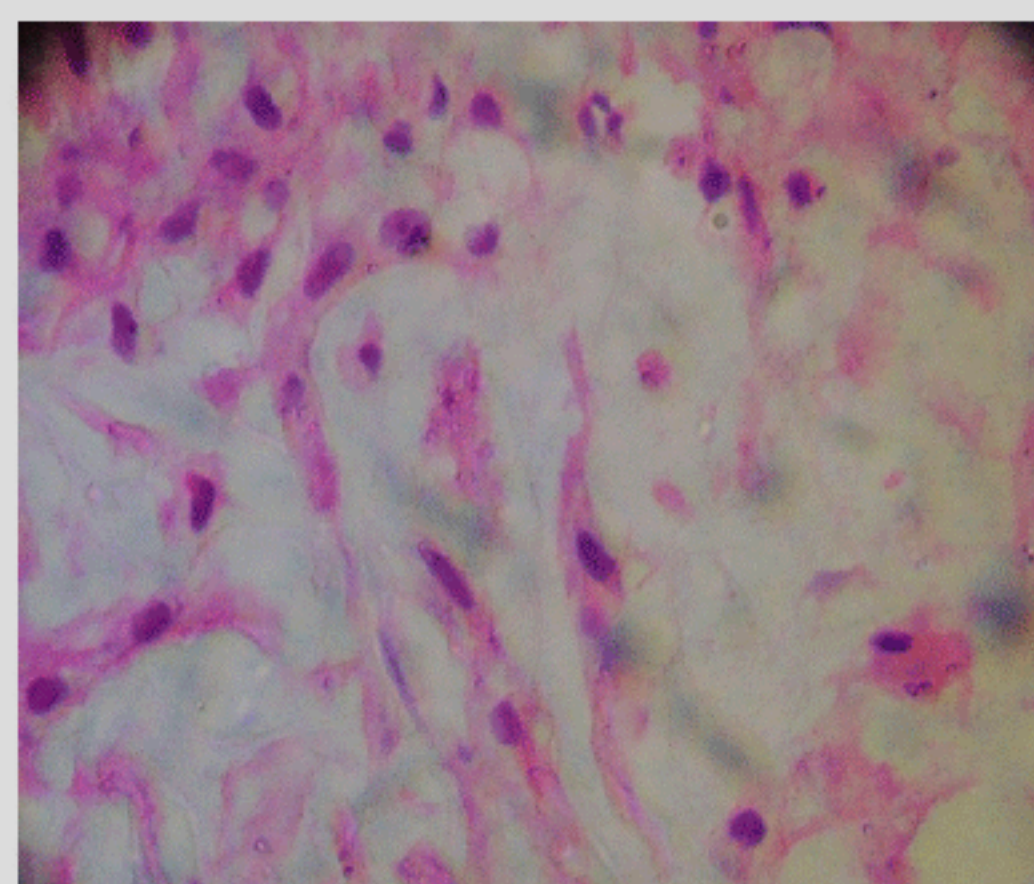


Figure 2 : Myxoid background of chordoma (HE x40)

1. Specialist - Histopathology, Department of Lab Medicine, Apollo Hospitals Dhaka, 2. Registrar, Department of Neurosurgery, Apollo Hospitals Dhaka, 3. Senior Consultant and Co-ordinator, Department of Neurosurgery, Apollo Hospitals Dhaka, 4. Senior Consultant and Co-ordinator - Histopathology, Department of Lab Medicine, Apollo Hospitals Dhaka

Chordoma

noted (Figure 1 & Figure 2). Histomorphologically, the tumour was diagnosed as chordoma.

Discussion: Chordomas are relatively rare neoplasms that appears to originate from benign remnants of primitive notochord. They account for about 0.15% of all intracranial neoplasm³. Kakuno Y has reported a case of 75 year old male⁴, Thoudou described three cases which was mimicking pituitary adenoma⁵. Watlkin's and his team reviewed 38 cases of skull based chordomas treated at the National hospital for Neurology and Neurosurgery in U.K. from 1958 to 1988. In their study, they found mean age of patient 44.3 years, male : female ratio was 6 : 5 with commonest presentation was cranial nerve palsy (94%) and headache (60%)⁶, a finding similar to our case.

The natural history of chordoma is a slow local growth and local invasion².

The best treatment is wide surgical excision followed by high dose radiation. The recurrence rate ranges from 28 %

to 80% cases and mean rate of metastases 30%². Prognostic factors include extent of resection, patient age (< 40 do better) and presence of mitotic activity¹.

References:

1. Burger CP, Scheithauer WB, editors, Tumours of the central nervous systems: AFIP fascicles: 1994. p. 303-306.
2. Forest M, Tomeno B, Vanel D, editors, Orthopedic surgical pathology, Philadelphia: Churchill Livingstone; 1988. p. 397-413.
3. Haridas A, Ansari S, Afshar F. Chordoma presenting as pseudoprolactinoma. Br. J Neurosurg. 2003;17(7):260-2.
4. Kakuno Y, Yamada T, Hirano H, Mori H. Chordoma In the sella turcica. Neurol Med Chir (Tokoyo). 2002;42(7):305-8.
5. Thodou E, Kontageorgos G, Scheithauer BW, Lekka I, Tzanis S, Mariatos P. Intracellular chordomas mimicking pituitary adenoma. J Neurosurg. 2000;92(6):976-82.
6. Watlkins L, Khudados ES, Kaleoglu M, Reves T, Crockard HA. Skull base chordomas: a review of 38 patients. 1958 - 88. Br. J Neurosug. 1993;7(3):241-8.