Nasopharyngeal Chordoma

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NASOPHARYNGEAL CHORDOMA is a rare but distinct entity. The terms ecchondrosis sphenoccipitalis, ecchondrosis physalifora and chordoepithelioma are synonymous.

The purpose of this paper is to present a case with uncommon clinical features and discuss this condition and consider the differential diagnosis.

Case Report

S.H., an eight-year-old Malay female, complained of progressive nasal obstruction worse on the left side for six months. The nasal obstruction had got more severe in the last month. She also complained of mucopurulent nasal discharge which was stained with blood when she blew her nose hard. There was anosmia in the left nasal cavity. There was no history of headache, diplopia or loss of weight. The past and family histories were non-contributory and her general examination and routine laboratory investigations did not reveal any abnormality.

Clinical examination revealed that the nose was expanded with splaying of the nasal bridge. There was also obliteration of the left naso-labial furrow (Fig. 1). Anterior rhinoscopy showed that the cartilaginous part of the nasal septum was pushed to the right side. Mucopurulent discharge was seen in the floor of the left nasal cavity. There was an irregular mass completely filling the left nasal cavity. This mass was greyish-red in colour and bled easily on manipulation and did not shrink when sprayed with 10% cocaine solution. The right nasal cavity was narrowed. Examination of the post nasal space showed that there was a mass filling the left posterior choana and protruding slightly into the nasopharynx.

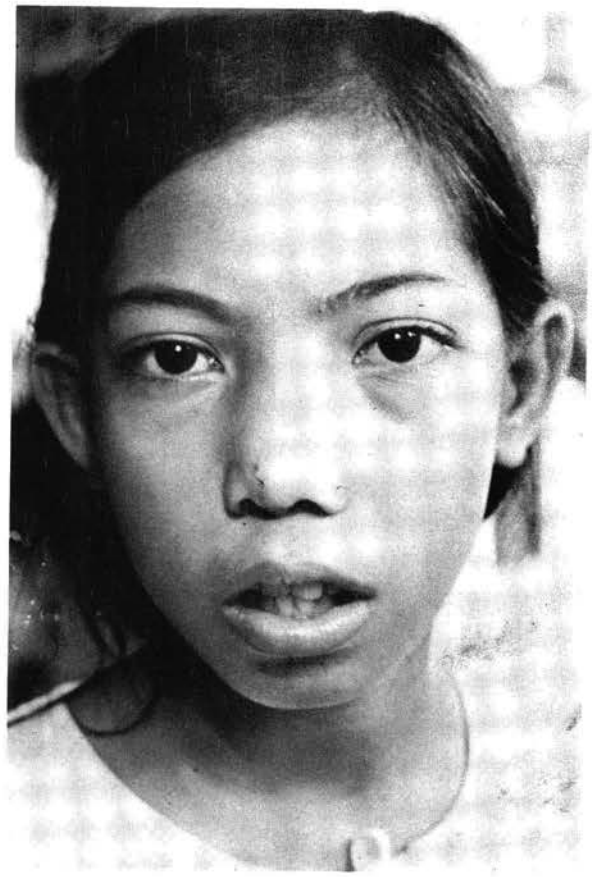

Fig. 1
Front view of patient showing splaying of the nasal bridge and obliteration of the left naso-labial furrow.

A smaller mass of adenoids was seen in the nasopharynx. Examination of the ears, throat and neck were normal. There was no sign of intracranial extension and eye signs were conspicuous by their absence.

Radiological examination showed a large soft-tissue mass in the nasal cavity with destruction of the nasal septum, lateral wall and floor of the nasal cavity consistent with a space-occupying destructive lesion on the left side. The mass was seen to extend forward from the posterior choana. Another soft-tissue mass was seen in the post nasal space consistent with enlarged adenoids. The left maxillary sinus was uniformly opaque (Figs. 2, 3 & 4).

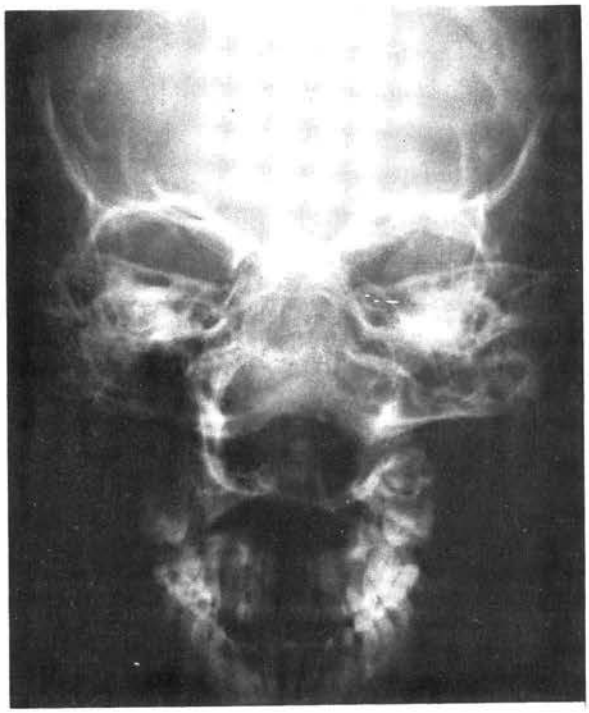

Fig. 2
Showing destruction of the Nasal Septum.

In view of the fact that the lesion tended to bleed easily, the patient was given a general anaesthetic and the pharynx was packed. An intravenous drip was started. The lesion in the left nasal cavity was soft and bled easily. The lesion appeared to be arising from the posterior part of the roof of the left nasal cavity and protruding through the left posterior choana. A portion was removed for histopathology. The portion that was removed appeared like jelly and felt gritty in parts. A BIPP pack was put in the left nasal cavity to arrest haemorrhage.

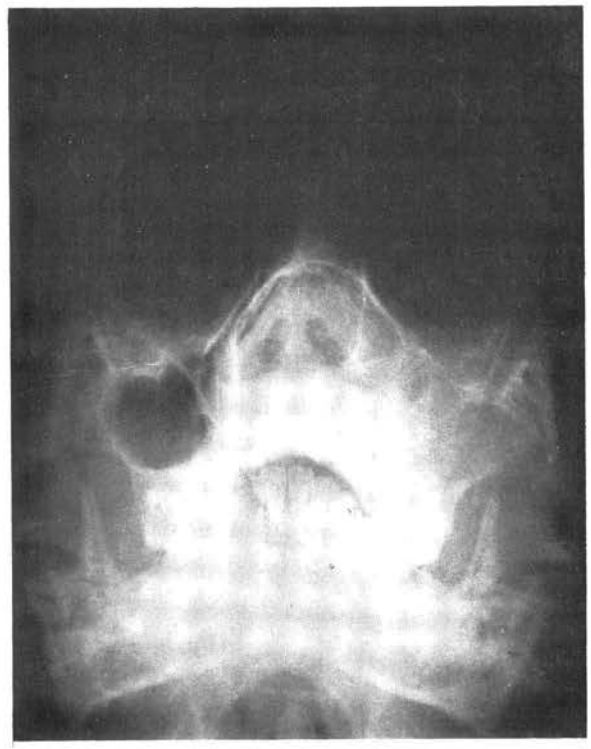

Fig. 3
Showing expansion of nasal cavity together with destruction of lateral wall of Maxillary Sinus. The left antrum is opaque due to secondary sinusitis.

Histopathological report (Figs. 5, 6 & 7).

The specimen consists of small fragments of soft gelatinous tissue and bone fragments. Microscopically, the tumour was composed of myxoid and mucoid stroma in which spindle-shaped and stellate cells were present. Also seen were several "physaliphorous cells", that is, large cells with vacuolated, bubble-like cytoplasm. There was also evidence of bone invasion by the tumour. The appearances are consistent with a chordoma.

The patient refused any form of treatment and went home.

Discussion

A chordoma is a tumour developing from embryonic remains of notochord. Hence, these tumours may arise anywhere along the axis of the notochord. Occasionally, fragments of the notochord become isolated, then chordomas may be found in unusual positions such as the alveolar process of the mandible, the maxilla, tonsillar region and superior portion of the occipital bone (Hass, 1934).

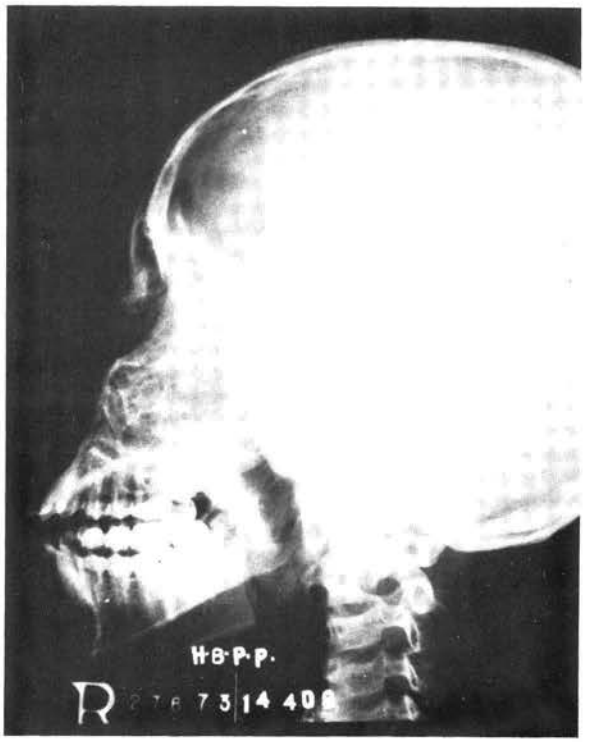

Fig. 4
Showing the chordoma in the nasal cavity and posterior choana. Also seen is a mass of adenoids.

Chordomas are usually more common in the sacro-coccygeal region than the cranial region and are three times more common in men than women. The incidence is less than one per cent of all central nervous system tumours (Poppen and King, 1952). The tumours are soft mucinous, slow growing but infiltrative and malignant. They are often large at the time of diagnosis. The histological picture is of an epithelioid or sarcomatous general architecture with a characteristic large vacuolated physaliferous cell containing an abundance of glycogen (Anderson, 1966). Local recurrence after surgical removal of this tumour is common. The tumour rarely metastasizes to lymph glands, liver, lungs, thyroid and skin.

The clinical picture of Cranial Chordomas depends largely on the direction in which the growth extends. Focal symptoms of the central nervous system owing to involvement of structures at the base of the brain are the usual presenting symptoms. In these cases, headache is a common symptom. The cranial nerves are commonly involved. The tumour may bulge into the cerebello-pontine angle and produce facial paralysis, unsteadiness, sensorineural deafness and loss of corneal reflex with numbness of face on the corresponding side.

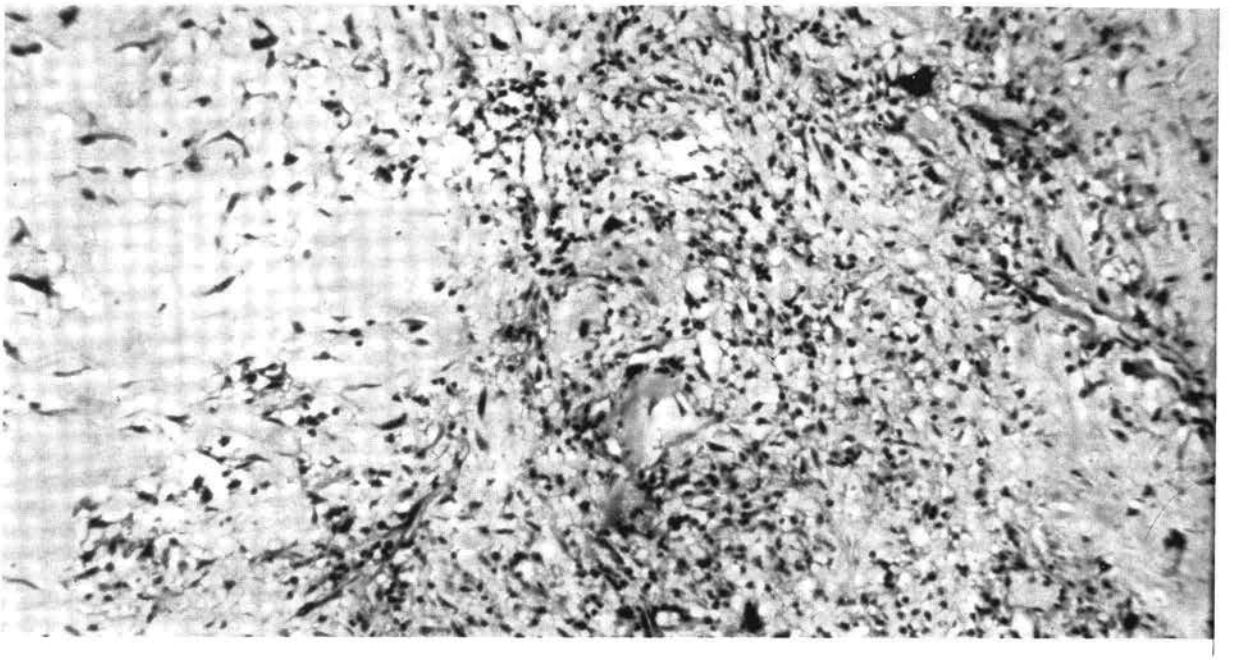

Fig. 5 *Photomicrograph*

Nasopharyngeal Chordoma **(× 100) – General appearance of the tumour showing many stellate cells and some physaliphorous cells interspersed within a myxoid stroma.**

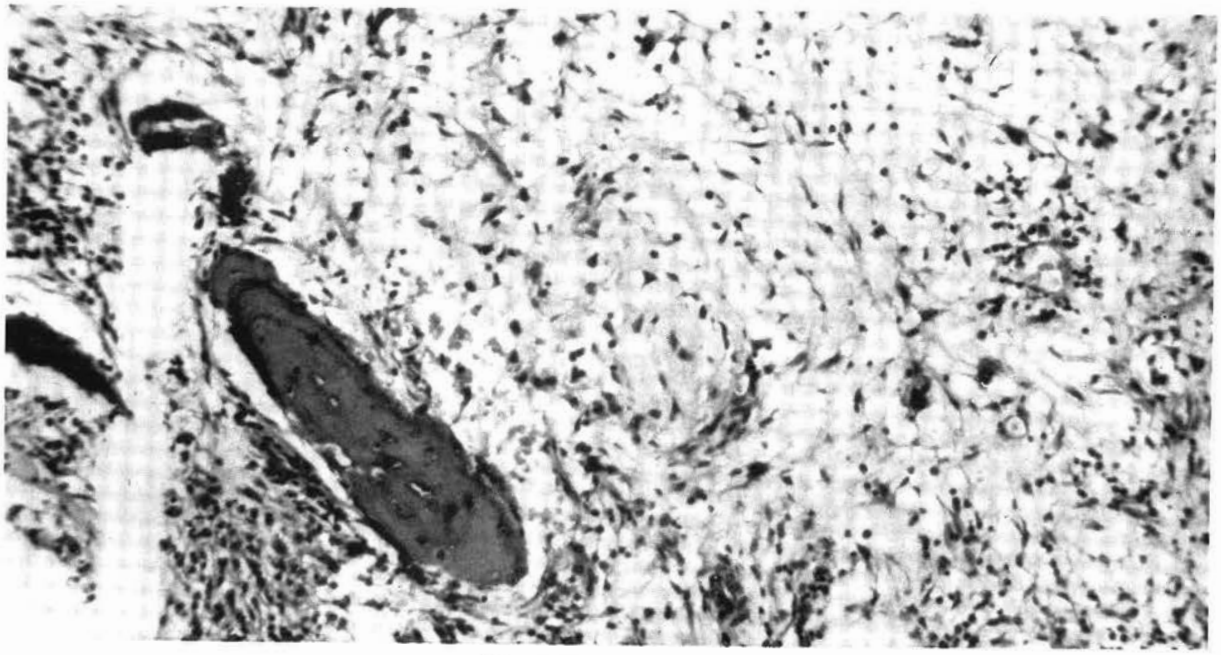

Fig. 6 *Photomicrograph*
Nasopharyngeal Chordoma (× 100) – **showing an area in which there is invasion of bone.**

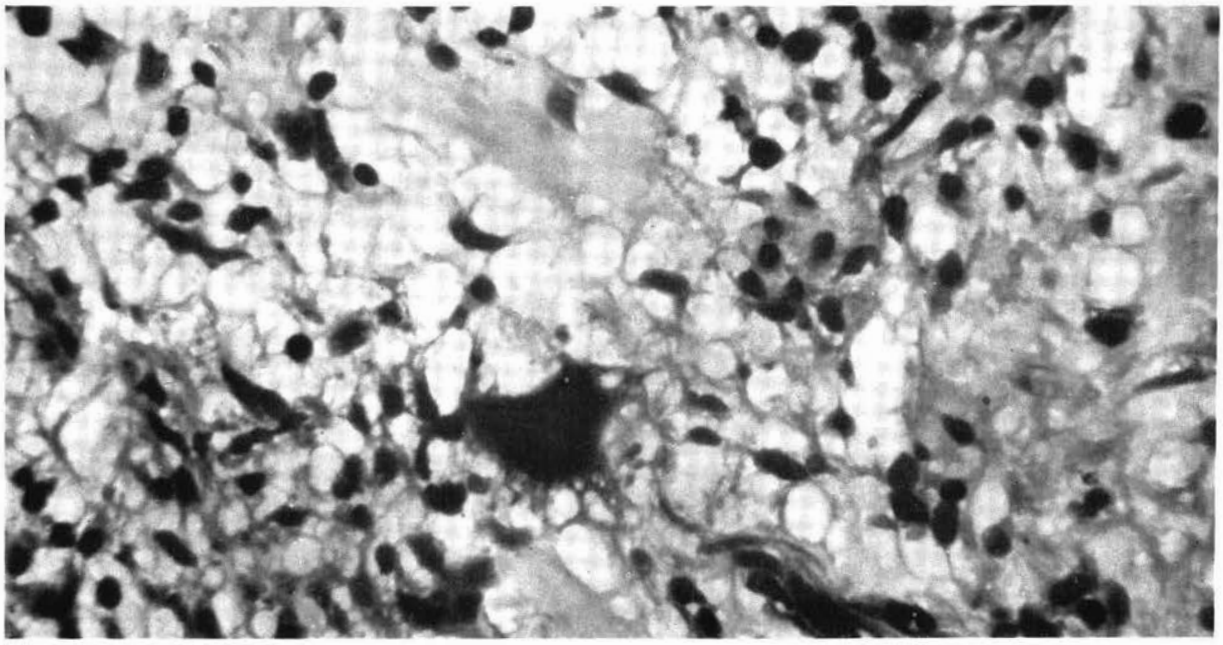

Fig. 7 *Photomicrograph*
Nasopharyngeal Chordoma (× 400) – **showing physaliphorous cells with their characteristic vacuolated ("bubble-like") cytoplasm.**

Visua symptoms are very common. In a comprehensive review of the literature on chordomas, GIVNER (1945) stressed the ophthalmological findings. There may be visual impairment and episodic diplopia. The lateral rectus muscle is frequently paralysed due to involvement of the abducent nerve. Less commonly, the oculomotor nerve is involved with corresponding paralysis of the muscles supplied by it. Visual field defects occur and there may be papilloedema.

Involvement of the pituitary gland is uncommon and evidence of pituitary insufficientcy is unusual and usually confined to females.

The tumour may grow into the nasopharynx. In these cases, there will be nasal obstruction. Infection is common and mucopurulent discharge which may be tinged with blood may occur. The infection may spread to the sinuses.

The orifices of the eustachian tubes may be obstructed and the patient may complain of tinnitus and deafness. The tympanic membrane is retracted and dull and its mobility is sluggish. Occasionally, a fluid level may be seen. The type of deafness in these cases is conductive.

Radiological study is helpful in the diagnosis of nasopharyngeal chordomas. Almost invariably, there is evidence of erosion of bone. When intracranial signs are present, ventriculography may be helpful. The final diagnosis is made on histology.

The case presented here is distinct because of the uncommon clinical features. The symptoms and signs were confined to the nose and nasopharynx. The patient is much younger than most of the patients reported. Only one other case below the age of ten has been reported in the literature.

In the differential diagnosis, the nasopharyngeal chordoma should be distinguished from juvenile angiofibroma and nasopharyngeal carcinoma.

Juvenile angiofibroma occurs in the younger age group and has a lobulated appearance with fine vessels coursing over its surface. It is firm in consistency. There is greater tendency to repeated severe epistaxis in juvenile angiofibroma. A carotid angiogram shows a dilated maxillary artery with the characteristic flush in the tumour. Bone erosion is very rare in juvenile angiofibroma.

Although progression of symptoms may be identical with that of nasopharyngeal carcinoma, lymphatic metastases are early and common in nasopharyngeal carcinoma but rare in nasopharyngeal chordoma.

The treatment of nasopharyngeal chordoma has been discouraging. The location of the neoplasm makes complete surgical extirpation a virtual impossibility. Irradiation appears to have beneficial effects in some cases.

Summary

A case of nasopharyngeal chordoma with uncommon clinical features in a young girl of eight years is reported. The condition is discussed and the differential diagnosis is considered.

References

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