Clinical records

Hamartoma of nasopharynx

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Though the histopathology of group of motley masses, termed 'Hamartoma', is now fairly clear, these enigmatic developmental masses still baffle an unwary clinician and pathologist with their bizarre features, infrequent occurrence and not too uncommon mimicry of tumour-like growths.

The term 'Hamartoma' was first introduced by Albrecht (1904) signifying some sort of an error (hamartia—error) in the growth of any of the three germinal layers. Hamartomas are tumour-like, but non-neoplastic, malformations or inborn errors of tissue development, characterized by an abnormal mixture of tissues indigenous to the part with excess of one or more of these. But Willis (1953) cautions that this term should be used only when there is clearcut evidence of a developmental anomaly. The term 'malformations' means not only errors of development since birth, but also those due to disturbances of still immature tissues in the postnatal period as well.

Discussing hamartomas, Eichel and Hallberg (1966) pointed out their basic differences from teratomas and dermoids. The term 'teratoma' implies a spontaneous, autonomous, new growth derived from pluripotential tissue. It is foreign to the region where it occurs and is composed of all three germinal layers. Whereas, 'dermoids' are cystic neoplasms and originate from two germinal layers—ectoderm and mesoderm, but with ectoderm predominating. In earlier literature teratomas and hamartomas were included under the common name 'teratoids' concealing the modern view that the group contains two distinct pathological entities (Wright, 1958). On the one hand, the teratomas, are true neoplasm composed of 'multiple tissues of kinds, foreign to the part in which they arise' (Willis 1967) occurring as a range of structure of increasing complexity and size, and are seldom encountered except in association with multiple congenital abnormalities in still birth foetuses. On the other hand are hamartomas, simple congenital malformations composed 'exclusively of components derivable from the local tissue' (Willis 1968). They are spontaneous growths which produce an excessive number of cells that reach maturity and then cease to reproduce, so that the growths are self-limiting. In many cases the resulting lesions seem to represent a simple exaggeration of a normal physiological process (Boyd, 1961).

Hamartomas common to a specific germinal layer are the sebaceous adenoma of ectoderm, haemangiomas and certain fibromas of mesoderm and congenital intestinal polypi of endoderm. To add to the clinician's confusion, while most malformations are easily distinguished from tumours, a hamartoma more often than not presents many clinical features of a tumour though basically it is a malformation. Hamartomas arise in many organs and tissues and inside mostly

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angiomas and benign pigmented naevi. Many of them do not have the ability of continuous growth and therefore can only be considered to be 'tumour like malformation'. There are others, however, that do show progressive proliferation, a basic character of a true tumour (Anderson, 1966).

Hamartomas of head and neck

In reviewing the early literature we find a small number of case reports of hamartomas involving the head and neck region.

Evans (1956) wrote on hamartomas of cartilaginous type in larynx, trachae, bronchi and lungs. Fuller (1963) reported pedunculated hamartoma of the oesophagus. Climie *et al.* (1964) described a lymphoid hamartoma of the larynx arising from the right ventricle. Perri (1956) and Ishii *et al.* (1968) described such malformations in the tongue. Eichel and Hallberg (1966) reported a case where a rare type of hamartomatous growth originating in the eustachian tube and filling the middle ear was removed surgically. From India, Kacker and Dasgupta (1973) reported hamartomas of the ear and nose and failed to find any previous reported case involving the nose or paranasal sinuses in the literature.

The early literature concerning these rare congenital tumours including one of Nasopharynx was reviewed by Kelly (1918) and later by Ehrich (1945), who found altogether 73 cases of hamartoma of different areas under the common nomenclature of 'teratoids'. Birt and Jones (1969) reported a case of nasopharyngeal hamartoma in an infant.

Hamartomas are commoner than teratomas and occur as hairy polypi of nasopharynx (Foxwell and Kelham, 1958; Bicknell, 1967). Hairy polypi may cause acute respiratory obstruction immediately after birth or feeding difficulties and cyanotic attacks over a period before they are discovered. Rarely they are an incidental finding in the adult. In our present case the hamartoma was found to be involving the nasopharynx and choanae causing nasal obstruction and it was removed by a transpalatel approach.

Case report

A 19-year-old male, R. (H. No. 689347), attended JIPMER Hospital on 30.11.1973 with complaints of nasal obstruction of a few months duration along with right sided otorrhoea. On examination his general build and health were found to be within normal limits. On otolaryngological examination he was found to be suffering from right otitis media with central perforation and mild conductive deafness; chronic rhinosinusitis with seromucinous nasal discharge (both sides); and a nasopharyngeal mass obstructing the posterior choanae, but more on right side, having the look and boggy feel of an antrochoanal polyp. In addition, there was a slight bluish discoloration over the hard palate (more on the left side) in the midline and near the posterior end. X-ray of the paranasal sinuses, suggested mild mucosal haziness of the maxillary sinuses more on the right side; routine blood, urine, and other laboratory investigations revealed nil abnormal. B.P., Pulse and Temp.—were within normal limits.

Initially, a provisional diagnosis of a big antrochoanal polyp with rhinosinusitis and c.s.o.m. right was made. On 4.12.1973 under general anaesthesia, removal of the antrochoanal polyp was attempted. In the operation, the other-

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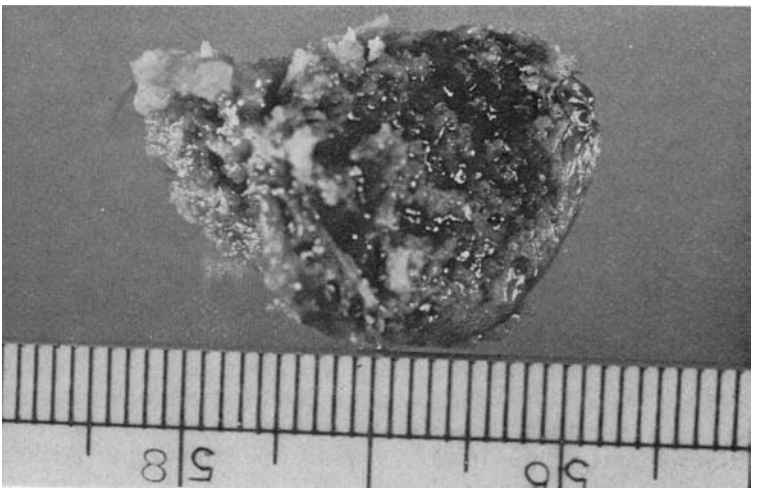


FIG. 1.

Photo of specimen removed. Picture of the hamartoma removed en mass through trans-palatal incision from nasopharynx. Solid mass of tissues with infiltration into bones seen.

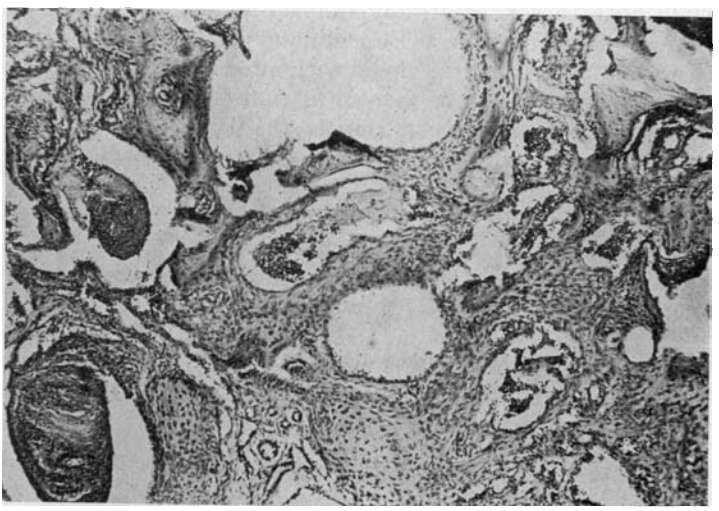


FIG. 2.

Microphotograph of hamartoma showing fibrovascular tissue with dilated and congested vascular channels with areas of cartilaginous tissue under-going ossification, in between them. The features characterizing Angiomatous Hamartoma.

wise simple manouvre of polyp avulsion seemed to be very tough because of an unusually firm feel, absence of a definite stalk, and associated massive haemorrhage from the mass. Piecemeal removal of good bits of tissues was done in an attempt to open up a fair amount of nasal passage which appeared completely blocked on the right side, with only a little chink still present at the left choanal aperture. The mass was found to be occupying the posterior 1/3rd of both nasal

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chambers and part of the nasopharyngeal vault, extending to the lateral walls —almost occluding the right eustachian tube orifice and causing thereby chronic tubotympanitis and otitis media. At this stage, due to the unexpected pathology, lack of preparation and associated haemorrhage, we decided to postpone the operation after placing in situ tight anterior and posterior nasal packs.

The immediate post-operative period was rather uneventful but the histopathological report of the bits removed came as 'angiomatous hamartoma (mostly cavernous spaces) infiltrating both soft tissues and bones'—on 10.12.1973.

On the basis of the histopathological findings and further clinical evaluation, the true nature of this bizarre mass and its probable extent became clear to us. We could understand why there was a bluish tinge over the mucosa on the oral aspect of the hard palate. The mass itself was found to be irregular, covering a wide area of the posterior part of the nasal chamber and nasopharynx.

So on 15.1.1974, under general anaesthesia, the mass was explored by Wilson's transpalatal approach. It was seen to be infiltrating the posterior 1/4 of the hard palate on the left side along with the soft tissues and adjoining bones. The mass was removed in bits by chisels and punch forceps, with a thin rim of normal tissues all around. At the end of the operation, to maintain post operative patency, a rubber tube of appropriate size was kept in either choana. Haemorrhage, though brisk earlier, practically stopped with complete removal of the hamartomatous mass. However, both post nasal and anterior nasal packs were inserted. The palatal incision was closed with interrupted 2/00 silk sutures.

The postoperative period was uneventful and the patient was reviewed at regular intervals for the next year. But for the persisting otitis media, there were neither any nasal complaints nor any local evidence of recurrence.

Summary

The extreme rarity of nasal and nasopharyngeal hamartoma, and its possible confusion in early diagnosis, prompted us to undertake a brief review of hamartomatous masses in E.N.T. practice along with this case report.

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