Clinical Records

Fibromatosis on the dorsum of the nose

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Abstract
An extremely rare case of fibromatosis in a 25-year-old Indian male is reported. The clinical examination and pre-operative findings were suggestive of a benign neoplasm, probably of neurogenic origin. The mass was well circumscribed and could be shelled out en masse. Histopathological examination showed it to be fibromatosis. An external rhinoplasty approach enabled a complete wide excision to be performed without any resulting cosmetic defect.

Introduction
Fibromatosis of the head and neck region refers to a controversial poorly defined group of cellular aggressive fibroblastic proliferative lesions that although benign may cause great difficulty in diagnosis and treatment. It is a rare tumour. It is especially rare in the nose and a similar lesion on the dorsum of the nose has never been reported in the literature. The following case report focuses on one of these rare tumours on the dorsum of the nose itself and how an external rhinoplasty approach enabled a complete removal of the mass with a cosmetically acceptable nose.

Case report
A 25-year-old Indian male presented with a four month history of a swelling on the dorsum of the nose. It had gradually increased in size without any pain or difficulty in breathing. There was no history of trauma or surgery to the nose. Clinical examination showed a 2x2 cm, well circumscribed, mobile, non-tender, firm swelling over the dorsum of the nose. The skin over the swelling was normal (Fig. 1). A lateral X-ray of the nose showed that the underlying bone and cartilage were not infiltrated. The swelling was removed en masse by an external rhinoplasty approach under a general anaesthetic. The post-operative appearance of the nose was cosmetically acceptable (Fig. 2). There was no definitive attachment to the underlying bone or cartilage. The post-operative period was uneventful.

The excised specimen consisted of an oval, grey-white mass (Fig. 3). The cut section showed a whistful fascicular surface over a slightly translucent greyish background. Histology showed spindle shaped cells arranged in fascicles. Abundant collagen was seen in between the tumour cells. Mitotic figures were few; nuclear atypia was absent. Adjoining muscles were infiltrated (Fig. 4).

Discussion
Fibromatosis or desmoid tumour is a benign fibroblastic proliferative lesion with abundant collagenous neo-formation located principally in the abdominal muscles and in the upper and lower extremities (Masson and Soule, 1966). It is a locally aggressive fibrous tissue tumour, generally does not metastasize, but may cause considerable morbidity and even death due to local infiltration. These are usually classified as fibromatosis rather than well differentiated fibrosarcoma, in order to indicate to the clinician that the tumour is a locally aggressive but not a metastasizing neoplasm (Fu and Perzin, 1976). Though uncommon, the common sites described include the retroperitoneal region, omentum, mesentry, small intestines, iliac fossa and breast. The incidence of these lesions in the head and neck varies from 9.5 to 12 per cent and out of them, 40-80 per cent of the tumours are located in the neck (Carlos et al., 1986). The maxillary and maxillary sinus, parotid, infratemporal space, parapharyngeal region, pharynx, larynx and lips are some of the rarer sites (Masson and Soule, 1966; Carlos et al., 1986).

Fibromatosis still poses difficult problems of diagnosis and treatment. It is frequently recurrent and infiltrates neighbouring structures. It is, hence, called an 'aggressive fibromatosis' or 'non-metastasizing well differentiated fibrosarcoma' (Fu and Perzin, 1976). Juvenile fibromatosis in the patients under the age group of 16 years is a variant and can involve paranasal sinuses along with gingival enlargement alone or as a part of systemic involvement (Naidu et al., 1991). As regards cause, little is known. It is attributed to trauma or alterations in the sex hormones (Carlos et al., 1986). The diagnosis is by histopathological examination: (1) The uniform growth, nature and appearance of fibroblasts and (2) the absence of both cellular atypia and mitotic figures favour the diagnosis.

The treatment is essentially surgical excision with a wide margin of adjacent uninvolved tissue. Radiotherapy, hormone treatment or chemotherapy are of no use (Wilkins et al., 1975; Majnudar and Winiarski, 1978). One exception is congenital or paediatric fibromatosis where chemotherapy can be useful (Naidu et al., 1991). Malignant transformation is extremely rare. The lesion usually grows rapidly but may take several years. Being benign, it does not metastasize. Infiltration of inoperable vital areas, after many years is the main cause of death.

This case has been reported for three important reasons:

1. Though intranasal or paranasal sinus involvement by fibromatosis has been reported in the literature, no desmoid tumour on the dorsum of the nose giving an external deformity has been reported in the English literature. There have been iso-
FIG. 1
Antero-posterior view of the nose. Note the swelling on the dorsum of the nose.

Radiated reports of cases involving the turbinates, maxillary sinus (Fu and Perzin, 1976) and sphenoid sinus (Carlos et al., 1986).

2. As described in the literature, an essentially benign lesion with histological infiltration into the surrounding tissues makes the clinical diagnosis difficult. There are no obvious or particular radiological findings attributable to fibromatosis. Hence, a clinical diagnosis of a benign neurogenic neoplasm was entertained pre-operatively.

FIG. 2
Post-operative photograph after three months. Note the cosmetically acceptable nose. There was slight hyperpigmentation on the site.

3. Surgical excision through an external rhinoplasty approach enabled a wide excision with no cosmetic defect on the most prominent and aesthetically significant part of the face—the nose.

Acknowledgements
The authors wish to thank Dr Ramdas M. Pai, Medical Director, Dr R. K. Rakshit, Medical Superintendent and Dr Rajamma Rajan, Director, Professor and Head of the Department of ENT, for permitting us to publish the paper and Miss Thulasi for secretarial assistance.

FIG. 3
The mass removed from the nose.

FIG. 4
Photomicrograph showing fascicles of spindle shaped tumour cells amidst which collagen is seen.
References

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Key words: Nose; Fibromatosis.