A lump on the nose

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DESCRIPTION

A young woman presented with prominence of the dorsum of her nose since childhood. She had become increasingly self-conscious of her appearance but was otherwise fit and well. She denied symptoms of nasal obstruction, epistaxis or discharge but described an intermittent ache related to the lump. There was no history of seizures, meningitis or cerebral abscess. Examination revealed a firm midline dorsal hump (figures 1 and 2). The rest of the examination was normal.

Nasal surgery was delayed into adulthood as not to impede nasal development. At the age of 19 years she underwent open septorhinoplasty for correction of what was thought to be a congenital osseocartilagenous deformity. A skin-coloured, fibrous mass of adipose tissue, skeletal muscle and nerve fibres was resected. This was histologically confirmed to be a subcutaneous hamartoma.

Hamartomas are benign tumours that represent the anomalous development of multiple tissue types native to the organ in which they are found.1 Most hamartomas are solitary, however multiple hamartomas may suggest tuberous sclerosis (associated with epilepsy), Cowden syndrome (macrocephaly, intestinal polyps, cutaneous hamartomas and increased cancer risk) or Proteus syndrome (overgrowth of skin, bones and other tissues).2

Systematic review of the literature found this to be the first documented case of an external nasal dorsum hamartoma. Nasal dorsum abnormalities are typically bony variations of normal nasal anatomy. The differential diagnosis for congenital midline nasal masses also includes dermoid cysts, gliomas and encephaloceles, which carry a risk of meningitis and cerebral abscess if they are found to contain intracranial connections.3

Learning points

▸ The differential diagnosis for congenital abnormalities of the nasal dorsum include bony variations in normal anatomy as well as dermoid cysts, gliomas, encephaloceles and, rarely, hamartomas.
▸ Hamartomas are benign tumours and are usually sporadic.
▸ Multiple hamartomas can be associated with heritable syndromes.

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Figure 1 Preoperative clinical photography demonstrates prominent nasal dorsum.

Figure 2 Anterolateral view.
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REFERENCES