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Nasal lipomas presenting as part of a syndromic diagnosis

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Abstract
An important part of the initial assessment of children presenting with congenital nasal masses is to exclude an intracranial extension using either magnetic resonance imaging (MRI) or computed tomography (CT) imaging.
We present three patients with such lesions in which unusual radiological findings were noted as part of the investigations.

Key words: Nasal neoplasms; Lipoma; Child; Tomography, X-ray computed; Magnetic resonance imaging

Case reports

Case 1

A two-day-old baby presented for investigation with a nasal mass protruding through the left nostril. Physical examination revealed a mass which appeared to be skin-covered. There was no associated rhinorrhoea and the remaining ENT examination was unremarkable. Associated with the mass were two other swellings, one on the forehead and one in the upper gingival sulcus (Figure 1). An MRI scan (Figure 2) was arranged to exclude an anterior cranial fossa defect. This, surprisingly, showed a lipoma in the corpus callosum and suggested that the forehead swelling was also a lipoma. Cardiac and renal ultrasound scans proved normal, as did ophthalmological examination. Under general anaesthesia, origin of the mass from the nasal septum was confirmed and it was excised, along with the gingival mass, leaving a patent nasal cavity. Pathological examination of the nasal specimen confirmed the diagnosis of lipoma.

Case 2

A three-month-old boy presented with a gradually enlarging swelling protruding from the right nostril (Figure 3), large enough to cause nasal obstruction and interrupt his feeding pattern. On examination, a skin-covered pedunculated swelling was seen to be arising from the nasal septum, protruding through the right nostril. To exclude an intracranial connection, a CT scan was arranged which did not show any abnormality. Formal excision of the mass was carried out under general anaesthesia via the naris since extension of mass along the septum was minimal. Histological examination confirmed it to be a lipoma.

Case 3

This case has been previously reported (Preece *et al.*, 1988). She presented at one month of age with a lesion protruding from the left nostril which had not changed since birth. Physical examination demonstrated a skin-covered lesion arising from the nasal septum blocking the nasal cavity. Also noted were two associated abnormalities, namely a notched upper lip and mesodermal dysgenesis of the anterior segment of the left eye (Figure 4). A

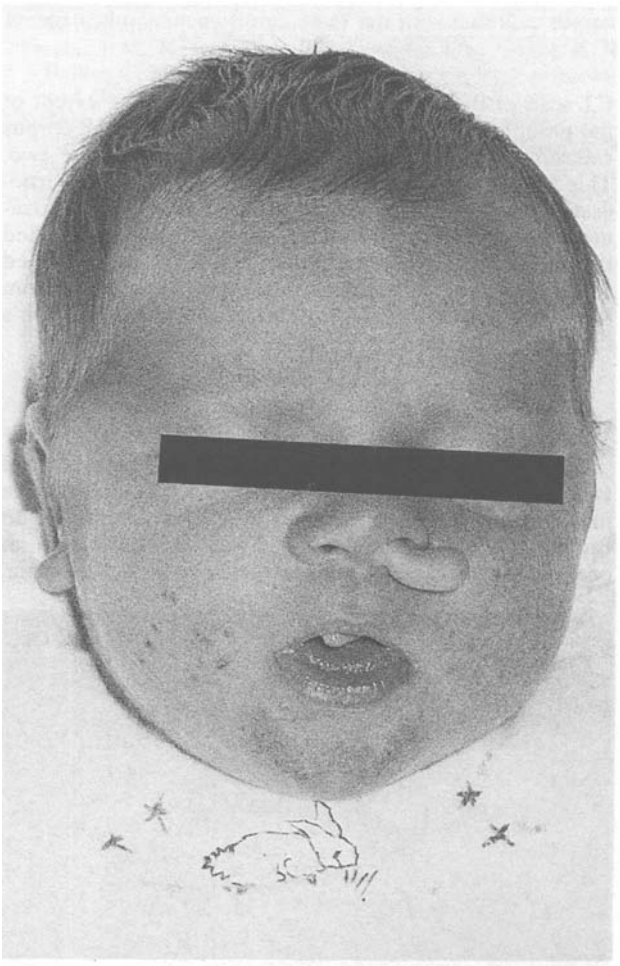

Fig. 1
Nasal and gingival lesions.

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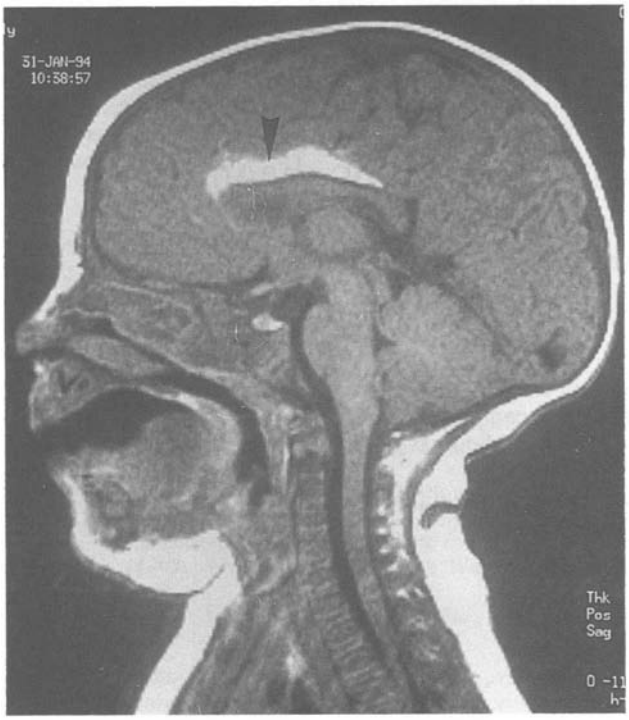

FIG. 2
Mid-sagittal T1-weighted MRI scan showing high signal in the corpus callosum with the same consistency as subcutaneous fat.

CT scan of the head (Figure 5) established the extent of the nasal lesion and demonstrated a lipoma of the corpus callosum with no obvious connection between the two. This finding was substantiated with an isohexol cisternogram. Initial treatment involved amputation of the extranasal portion of the lesion. Histological analysis verified the presumed diagnosis of lipoma. When the child reached 15 months of age a definitive resection of the lesion from the septal mucoperichondrium was achieved via a Weir's (alar base) incision.

Discussion

Midline nasal masses presenting in infancy are unusual and need to be accurately distinguished so that appropriate management can be initiated. It is most important to exclude an intracranial extension such as occurs with an encephalocoele. The differential diagnosis includes glioma,

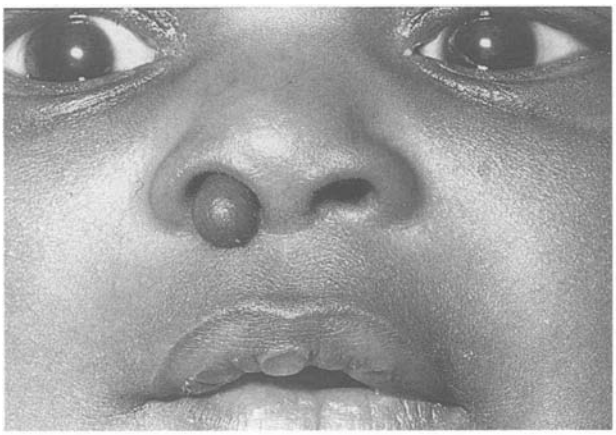

FIG. 3
Nasal lesion.

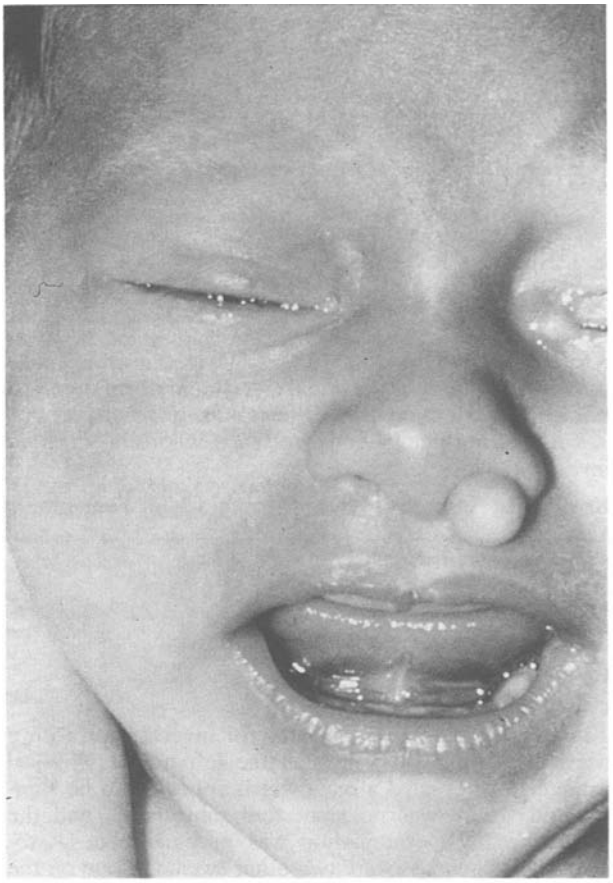

FIG. 4
Nasal lesion and notched upper lip.

nasal dermoid, teratoma, haemangioma and nasolacrimal duct cyst (Finkelhor, 1990). Lipomas are generally common but affect the head and neck with much less frequency than the rest of the body. Even more unusual are those which occur in the nasal cavity, paranasal sinuses and nasopharynx, presumably because adipose tissue in these areas is scarce (Batsakis, 1979). Lipomas affecting the central nervous system are even more infrequent, occurring with a frequency of 0.1 per cent, the majority of

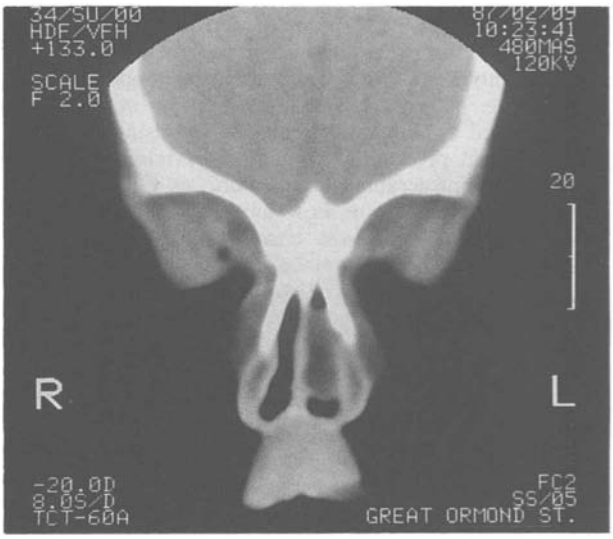

FIG. 5
Coronal section computerized tomogram demonstrating extent of nasal mass.

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which are in the corpus callosum (Graham, 1985). These lesions are usually asymptomatic but may be associated with seizures, raised intracranial pressure, dementia and hemiparesis (Tahmouresie *et al.*, 1979; Gerber and Plotkin, 1982). Surgical resection of a callosal lipoma is technically difficult due to the vascularity of the lesion and its often close association with the anterior cerebral arteries as well as adherence of the capsule to adjacent brain. Seizures, if occurring as a result of corpus callosal lipomas, can be treated with anti-convulsants (Kazner *et al.*, 1980).

The encephalocraniocutaneous lipomatosis syndrome (E.C.C.L.) is a rare disorder, first described in 1970, features of which include seizures, mental deficiency and unilateral cutaneous and ophthalmologic lesions with ipsilateral cerebral malformations. The exact pathogenesis remains unclear, but the defect appears to involve embryonic ectodermal and mesodermal elements at a critical period in development. As yet there is no clear suggestion of familial transmission. It is recognized that cutaneous and meningeal lipomas exist with lipomas of the central nervous system and viscera (Haberland and Perou, 1970). Fronto-nasal dysplasia is a disorder which exists as a spectrum of abnormalities in which median facial abnormalities occur together with ocular hypertelorism. The aetiology of the condition is unknown, no genetic basis having been found, although presumably it is due to interference with the normal embryological development of the face (Sedano *et al.*, 1970).

The three cases described illustrate a spectrum of disease. The first case illustrates a form of E.C.C.L. since a cutaneous lipoma co-existed with that of the central nervous system. Full expression of the syndrome was not evident, however, since seizures and mental deficiency were not apparent at the time of presentation. The second case was simply that of a septal lipoma with no other associated abnormality. At the time of presentation of the final case, it was not fully appreciated that a syndromic diagnosis might apply. With hindsight it is difficult to make an exact diagnosis since some features of both fronto-nasal dysplasia and E.C.C.L. existed. The median notched upper lip is suggestive of fronto-nasal dysplasia, but the cutaneous and central nervous system lipomas together with a unilateral ophthalmic lesion imply that E.C.C.L. is the correct diagnosis, particularly as hypertelorism was not evident.

These cases illustrate unusual features which may be found during the pre-operative assessment of infants presenting with congenital intranasal masses and emphasize the need for careful radiological investigation. This approach is necessary to prevent blind surgery being performed in the presence of an intracranial connection, which could lead to a CSF leak or meningitis. The possibility of an associated syndrome should also be borne in mind when other abnormalities are present and appropriate clinicians should be involved in the patient's assessment and management.

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