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Middle ear teratoma in a newborn

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Abstract
Teratomas are tumours that rarely exist in children. Their location in head and neck is, however, an unusual finding.

A strange case of middle ear congenital teratoma reaching the cervical area is presented here. In the last 20 years it is only the second case to be published in the English literature. The tumour was revealed by the presence of a peripheral congenital facial palsy.

Key words: Facial paralysis; Ear, middle; Teratoma; Abnormalities

Introduction

Teratomas are a variety of tumour derived from the three germinal layers (ectoderm, mesoderm and endoderm). Other wide definitions include dermoid cysts although these only contain elements from the ectoderm and mesoderm (Rothschild *et al.*, 1994).

The incidence of teratomas is approximately 1:4000 in live newborns (Forrest *et al.*, 1993). Malignancy, determined by immature cellular elements, is observed in 20 per cent of cases, being more common in adults (Grosfeld and Billmire, 1985). Although teratomas can appear in any part of the body, they usually appear in midline and para-axial locations. The more common location is at the sacrococcygeal level (40–60 per cent) (Bale *et al.*, 1975; Grosfeld and Billmire, 1985; Talmi *et al.*, 1988). Head and neck teratomas are infrequent and vary between two per cent and 10 per cent of cases according to published statistics (Grosfeld and Billmire, 1985; Jordan and Gauderer, 1988). There is no reference in the widest consulted series about the location of teratomas in the middle ear. One isolated case of a middle ear teratoma spreading to the nasopharynx was published in 1993 by Forrest *et al.* Its behaviour in the middle ear's air cavity is similar to the case presented.

This article describes a strange case of a middle ear teratoma reaching the cervical area in a live newborn infant with peripheral congenital facial palsy. The rareness of the case caused many diagnostic problems.

Case report

The patient is a male, born at 36 weeks through a non-instrumental delivery after a controlled pregnancy without any significant alterations. There was no antecedent of polyhydramnios. His mother, a native of Maghreb, had among her antecedents an infant who died on the third day of life, cause unknown.

The patient was admitted to our hospital in the second week of life, with a peripheral palsy of the seventh cranial nerve, Grade 5 in the House-Brackmann's classification. The analytical study and the usual viral indicators were normal. The serum values of alpha fetoprotein were 872 ng/ml. Cervical palpation revealed the presence of a high, soft, sunken, apparently cystic-looking laterocervical mass. The two possible diagnosis were branchial cyst and cystic lymphangioma; however, it was decided to perform magnetic resonance imaging (MRI) and a CT scan due to the convex nature of the tympanic membrane without any visible signs of inflammation.

The MRI revealed the existence of a solid-cystic mixed mass with calcareous deposits, to the side of the large neck vessels in the infraparotid space.

The CT scan demonstrated a mass filling the middle ear cavity and projecting into the Eustachian tube. The ossicular chain was apparently complete.

A middle ear tuberculosis or a congenital cholesteatoma was considered possible. The first possibility was rejected

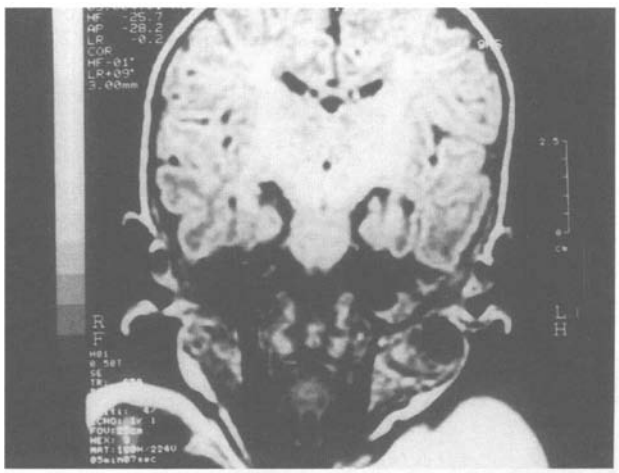

FIG. 1
Mixed solid and cystic mass in upper lateral region of the neck (MRI).

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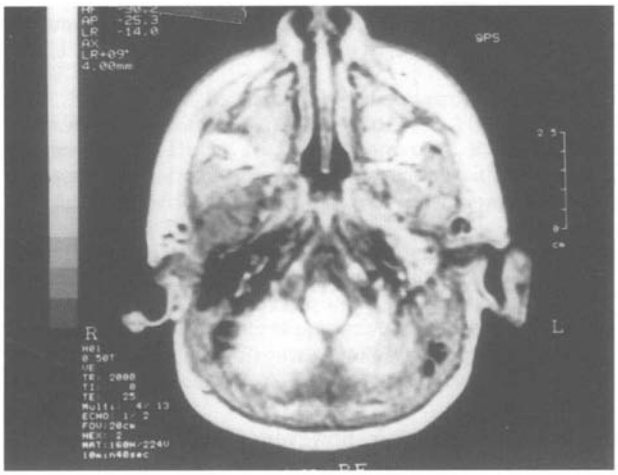

FIG. 2
Middle ear mass extending into the Eustachian tube (MRI).

due to the absence of clinical signs compatible with congenital tuberculosis and a negative Mantoux test. The cervical involvement made a cholesteatoma unlikely. The execution of a biospy through an exploratory tympanotomy was chosen because fine needle aspiration biopsy was negative. The presence of a first degree immature teratoma was confirmed.

The patient was operated using a combined retroauricular and cervical approach, a fibrous-looking soft mass was found in the middle ear. Some fibrous tracts connected the cervical mass to the middle ear teratoma through the mastoid, and compressed the facial nerve on its exit from the stylomastoid foramen.

After six months, the patient is free of disease.

Discussion

There are a great variety of teratomas classified according to their location, histological appearance and clinical behaviour. Their more common location is at the sacrococcigeal level, appearing in the neck in only three per cent of cases (Grosfeld and Billmire, 1985; Sanchez *et al.*, 1988; Olivan Gonzalvo *et al.*, 1989; Zerella and Finberg, 1990; Rothschild *et al.*, 1994). The malignancy of these tumours is determined by the immaturity of the cellular elements. This can be seen in approximately 20 per cent of cases. Most cervical teratomas in childhood are benign, unlike the ones in adults (Grosfeld and Billmire, 1985; Billmire and Grosfeld, 1986; Talmi *et al.*, 1988; Touran *et al.*, 1989). There have been fewer than ten cases with cervical lymph node metastasis and the presence of glial implants was proven in most of them (Gundry *et al.*, 1983; Tapper and Lack, 1983; Pearl *et al.*, 1986).

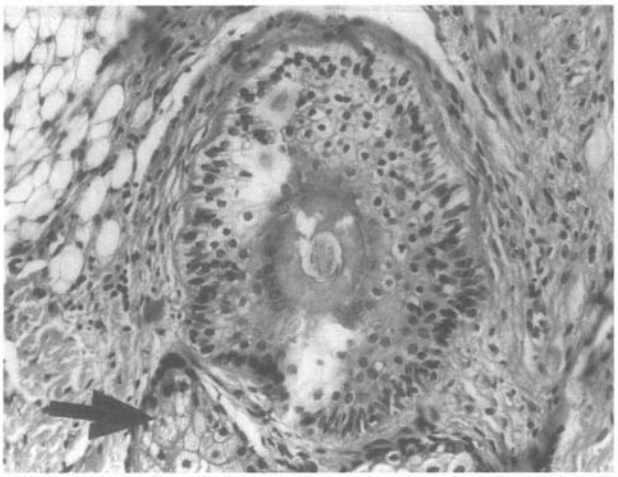

FIG. 3
Hair follicle, sebaceous gland (arrow), connective and adipose tissue (ME × 20).

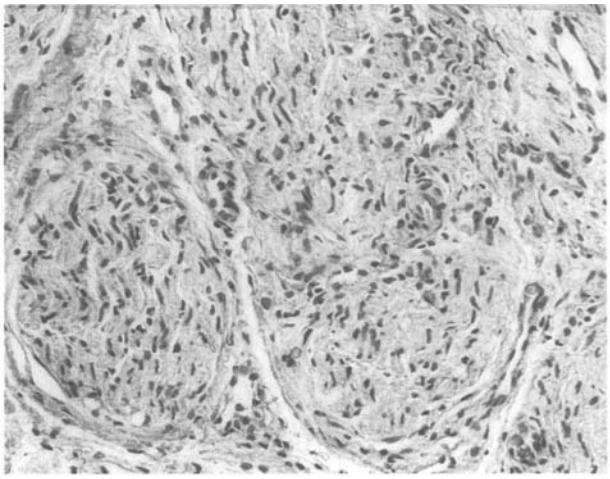

FIG. 4
Numerous nerve fascicles, transverse and oblique section (ME × 20).

Their histogenesis is not clear but all theories include the concept of an early event in embryogenesis that releases totipotent primitive cells, from which the tumour eventually arises. These cells go out of control during development, provoking different degrees of maturity in the different cell pathways.

The location of this neoplasm in the middle ear's air cavity is unusual. In the last 20 years, there has been only one reference to a middle ear teratoma spreading to the nasopharynx through the Eustachian tube (Forrest *et al.*, 1993). The patient also presented a cervical extension in the perimastoid region, responsible for the compression of the peripheral facial palsy. The behaviour of the tumour in the middle ear was similar to the one published by Forrest *et al.* (1993). The possibility of a congenital cholesteatoma was included in the initial diagnosis; however, this was questioned due to three characteristics:

1. no injury to the ossicular chain;
2. an important dilatation of the Eustachian tube entrance;
3. the cervical involvement of the tumour.

Conclusion

A congenital tumour in the middle ear is possible. A teratoma can occasionally exist in this area. The tumour does not usually involve the ossicles, tending to spread itself towards less resistant areas such as the Eustachian tube. An exploratory tympanotomy seems to be the most useful diagnostic method in dealing with this possibility. Early surgery with complete removal of the tumour is the most suitable treatment.

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