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Clinical Records

Unilateral hearing loss due to a rhabdomyoma in a six-year-old child

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Abstract
A case report of a six-year-old child is presented, who had had a unilateral sensorineural hearing loss for several years. Because of impairment in the ABR as well as in the caloric testing a MRI and CT scan were performed. A 17 mm tumour in the cerebellopontine angle (CPA) was detected, which after suboccipital surgery proved to be a rhabdomyoma. This tumour has not been described before in the CPA. Unilateral sensorineural hearing loss should, at all ages, be an indication for further (radiodiagnostic) investigations.

Key words: Rhabdomyoma; Hearing loss, unilateral; Cerebello-pontine angle; Child

Introduction

Unilateral sensorineural hearing loss and complete unilateral deafness seems to occur in one out of 1000 children of school age (Everberg, 1960). The cause most often remains unknown. Rarely a genetic origin can be traced (Marres, 1994). In adolescents and adults a small asymmetric sensorineural hearing loss requires an investigation of the cerebello-pontine angle to exclude a space-occupying lesion. A case report is presented of a six-year-old boy with a unilateral sensorineural hearing loss. After histological studies it was found to be a rhabdomyoma, which previously has been reported only once in an intracranial location in the trigeminal nerve (Zwick *et al.*, 1989). Nowadays, we have access to advanced imaging techniques of the inner ear and the cerebello-pontine angle, and modern radiology is non-expensive and comfortable for patients these additional investigations should also be considered in childhood.

Case report

In 1990, a six-year-old child was referred to the ORL department of the University Hospital, Nijmegen because of a unilateral hearing loss, found at a regular screening at school. There was no medical history. In a scheduled examination after the first year of life a hearing loss had been noticed, but no further action had been taken. The speech and motor development were normal.

At the ENT-examination a normal eardrum was seen, with air-containing middle ear. The tuning fork test according to Rinne and Weber were normal, and the Barany noise box test proved that both ears could hear. At pure tone audiometry a sensorineural hearing loss was found with a Fletcher Index (mean loss at 0.5, 1.0 and 2.0 kHz) of 50 dB in the left ear. Speech audiometry gave a word discrimination of 100 per cent with the same 50 dB shift. The hearing was normal in the right ear. Contralateral stapedial reflexes could be elicited on both sides. The ABR on the left side revealed an interwave delay from J1-J3 and J1-J5. On the right side a much smaller interwave delay J1-J5 was found. Labyrinth testing with calorics indicated a reduced function of the left labyrinth.

MRI scanning had just become available in a nearby hospital and a tumour of the eighth nerve was found (Figure 1). It was not possible to use Gadolinium at that time. Later, a CT scan with the use of contrast showed a tumour of 17 mm in the left cerebello-pontine angle (Figure 2). Angiography showed no abnormal vascular structures.

The patient was referred to the neurosurgical department, and a suboccipital approach was used for the exploration of the cerebello-pontine angle. During the operation the tumour showed encapsulation with a smooth surface and firm consistency. There was an ingrowth in a dilated internal acoustic meatus and adhesions were found with the pons. It was not possible to remove the tumour totally. Post-operatively there was a fast recovery, but a total facial paralysis House grade VI (House and Brackman, 1985) remained. The left ear was completely deaf. There were no vertigo problems afterwards. MRI-scanning three years after the operation showed no growth of the remaining part of the tumour. Four years after the operation, a cross face nerve transplant was made using the left sural nerve to reinnervate the left facial nerve.

Pathology

During the operation a frozen section examination was performed and because of the presence of matured skeletal muscle a diagnosis of teratoma was suggested. In the permanent paraffin sections the tumour showed large well-developed matured skeletal muscle fibres with easily identifiable haematoxylin and eosin stain cross striations, accentuated by PTAH-Mallory stain (Figure 3). The fibres

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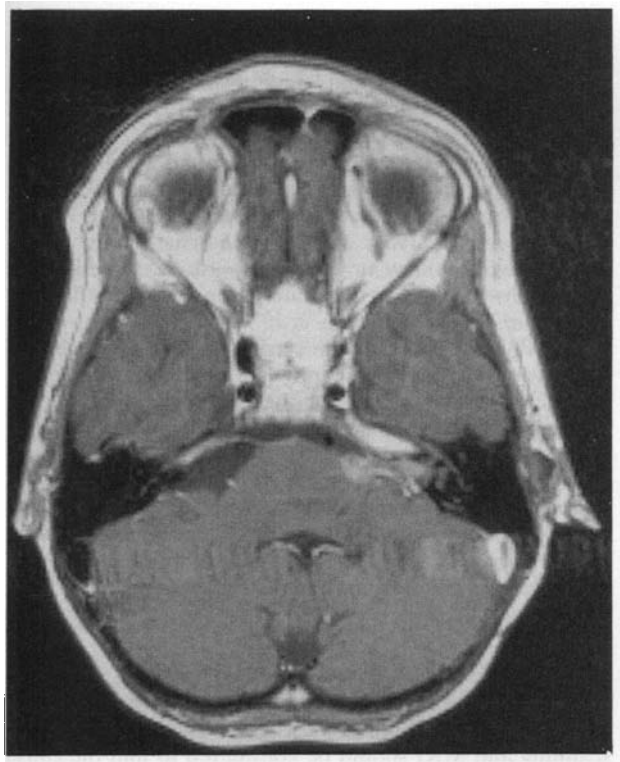

FIG. 1

MRI without Gadolinium of the rhabdomyoma in the left cerebello-pontine angle.

were arranged in irregular fascicles and separated by various amounts of fibrocollagenous tissue. No cystic structures, vacuolated cells or spider cells were found. There was no mitotic activity. Muscle fibres showed diffuse cytoplasmic immunoreactivity for desmin, muscle-specific actin HHF-35 and myoglobin. No glial tissue was detected and there was no reaction for S100 protein and GFAP. Some tumour cells stained also for vimentin.The diagnosis rhabdomyoma was made.

Discussion

Hearing loss and even deafness of one ear in childhood is a regular finding in daily practice. Usually, the hearing loss is due to middle ear problems and will be solved in time, by using medication or middle ear ventilation tubes. When a sensorineural hearing loss is found there is a serious indication for further investigations. Tumours as in this young child are rare but can exist for several years giving symptoms which should not be overlooked.

Congenital intracranial tumours are rare and seldomly found in the cerebello-pontine angle (Werb *et al.*, 1992). A rhabdomyoma is defined as a benign and usually circumscribed tumour consisting of mature muscle cells (Kleihues *et al.*, 1993). Other benign mesenchymal tumours with intracranial locations such as leiomyoma of fibrous xanthoma have also been reported (Burger and Scheithauer, 1994).

The location of rhabdomyoma can be cardiac or extracardiac. Cardiac rhabdomyomas are rare congenital tumours resulting from early dysembryogenetic disorder of organogenesis. They are generally benign lesions considered as hamartomas which may be the first manifestation of Bourneville's tuberous sclerosis (Mehta, 1993). The extracardiac rhabdomyomas may assume a number of histological patterns. Tumours composed of uniform large polygonal eosinophilic cells with occasionally cross striation are classified as the adult-type. Those consisting of small mesenchymal cells and spindled skeletal muscle cells are classified as the foetal-type. Both tend to occur in the head and neck region (Knowles and Jacobiec, 1975; Fu and Perzin, 1976; Kapadia *et al.*, 1993; Kleihues *et al.*, 1993). A third form, the genital-type with cells bearing resemblance to both the adult and foetal type is most often seen as a polypoid mass in the vagina and vulva (Agamaolis *et al.*, 1986; Enzinger and Weiss, 1988). Foetal extracardiac rhabdomyomas occur principally in infants and young children and have a predilection for the postauricular region (Batsakis and Manning, 1986) but are even rarer than the adult-type rhabdomyomas (Enzinger and Weiss, 1988). They show close resemblance to rhabdomyosarcomas. Of particular interest is their association with multiple basal cell carcinomas and anomalies of the rib and the iridocorneal angle of the eye (Enzinger and Weiss, 1988). In the adult form the tumour can be found multifocal in the head and neck region (Shemen *et al.*, 1992; Kapakia *et al* 1993). In the neck the adult type rhabdomyoma seems to arise from the branchial musculature of the third and fourth branchial arches. Therefore, the tumour is found most frequently in the region of the larynx, pharynx, floor of the mouth and base of the tongue (Solomon and Tolete-Velcek, 1975; Enzinger and Weiss, 1988). Both adult and foetal type rhabdomyomas are more common in males (Agamanolis *et al.*, 1986; Kapadia *et al.*, 1993). Recently a juvenile-type of rhabdomyoma has been described which appears as an intermediate form between the foetal- and adult-type (Nakheh *et al.*, 1991; Crotty *et al.*, 1993).

Microscopically intracranial rhabdomyomas must be distinguished from rhabdomyosarcomas, medullomyoblas-

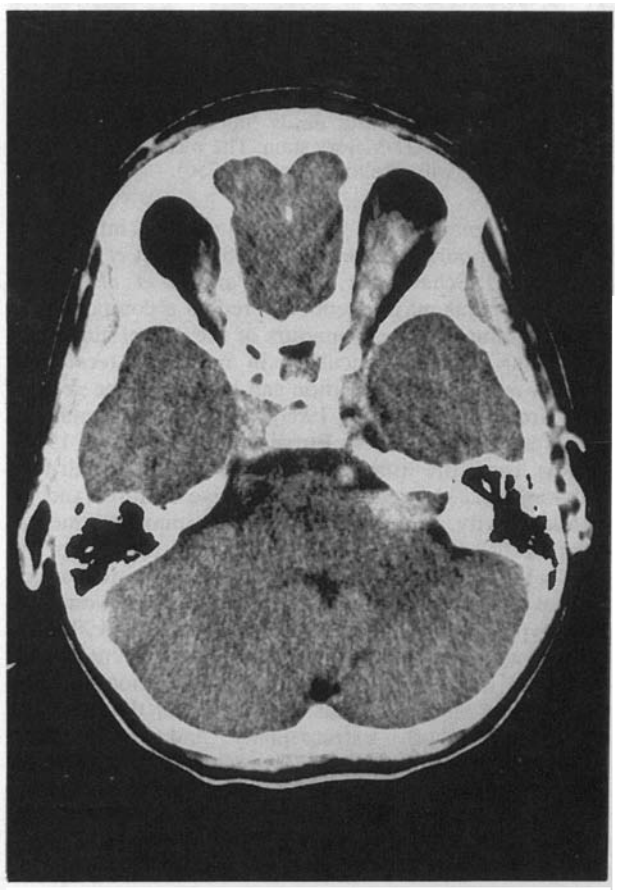

FIG. 2

CT scan with contrast of the rhabdomyoma in the left cerebello-pontine angle.

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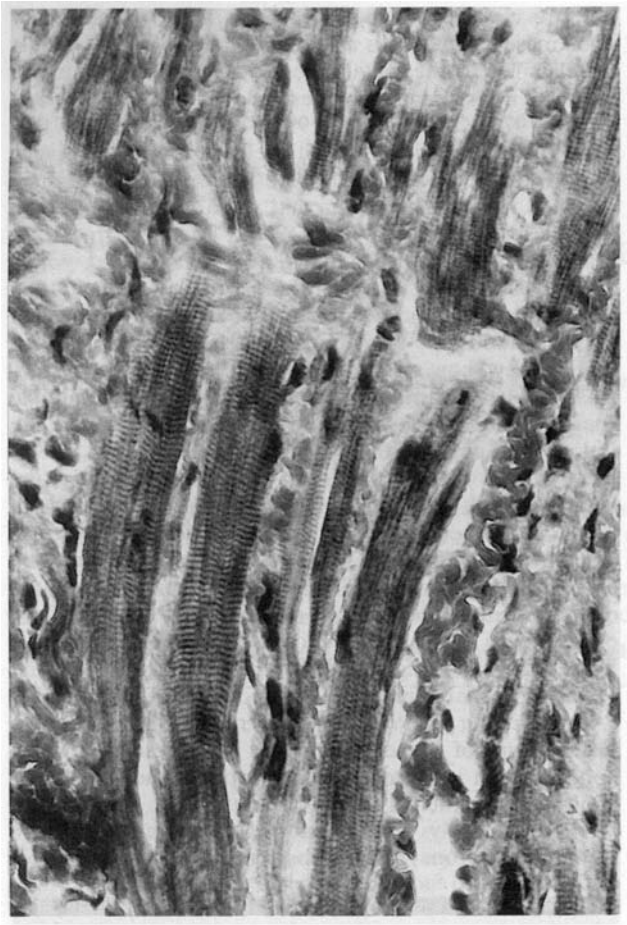

FIG. 3

Photomicrograph of the tumour illustrating well developed skeletal muscle fibres with easily identified cross striation accentuated by PTAH-Malory stain. The picture is consistent with rhabdomyoma (× 250).

tomas, malignant triton tumours and rare intracranial rhabdoid tumours (Jakate *et al.*, 1988; Kodet *et al.*, 1991; Burger and Scheithauer, 1994; Parham *et al.*, 1994). Rhabdomyomas may be encountered as a component of the malignant germ cell tumours of the pineal gland and suprasellar region, particularly in immature teratomas. Skeletal muscle differentiation can also rarely be seen in glycosarcomas (Goldman, 1969), gliomas and meningiomas (Ferracini *et al.*, 1982; Burger and Scheithauer, 1994). In this case the tumour classified as the adult-type rhabdomyoma showed other distinctive features and no mitotic activity. Immature neuroblasts, immature mesenchymal cells, vacuolated or globular rhabdomyoblasts and glial cells were not found. Our immuno-histochemical findings were consistent with the rhabdomyomatous phenotype (Helliwell *et al*, 1988; Kapadia *et al.*, 1993).

The four year follow-up time free of recurrent disease confirmed the benign character of the lesion. Overall incidence of rhabdomyomas is much lower than rhabdomyosarcomas both extracranially and intracranially (Enzinger and Weiss, 1988; Zwick *et al.*, 1989; Burger and Scheithauer, 1994). Rhabdomyosarcoma is a highly malignant tumour and patients stay alive no longer than two years despite aggressive chemotherapy and radiotherapy (Taratuto *et al.*, 1985).

It is a question of debate whether the presented case should be considered as a choristoma or a true neoplasm. We were able to find in the literature only one other case of a primary intracranial rhabdomyoma which involved the trigeminal nerve. The authors regarded the lesion as choristoma and postulated that the tumour arose from embryologically disrupted and dislocated masticatory myogenic tissue following innervation of the mandibular motor division of the trigeminal nerve (Zwick *et al.*, 1989). The foci of skeletal muscles can be found accidentally in normal meninges, but their relationship to rhabdomyomas and rhabdomyosarcomas is not clear (Hoffman and Rorke, 1971; Fix *et al.*, 1989; Burger and Scheithauer, 1994). Rhabdomyosarcomas have been reported to arise in both the leptomeninges and the brain parenchyma (Korinthenberg *et al.*, 1984; Taratutu *et al.*, 1985; Ferracini *et al.*, 1992;). Focal skeletal muscle ectopia in leptomeninges has been reported predominantly in children with other developmental anomalies of the central nervous system and sometimes chromosomal abnormalities which suggests faulty mesenchymal differentiation secondary to a genetic error in the tissue regulation. In several cases skeletal muscle has been found in leptomeninges of the pontine region or ponto-medullary junction (Johnson and Ludwin, 1984; Fix *et al.*, 1989). Interestingly, the tumour reported by us was also located in the pontine angle area.

Conclusion

Unilateral sensorineural hearing loss may be an alarming symptom in childhood. Further investigations using CT scanning and MRI should be considered to find the cause of the phenomenon. In childhood rare congenital tumours such as a rhabdomyoma are sometimes found in these cases and early exploration increases the chance of successful radical surgery. The presented child is alive and well four years after operation, although only an incomplete resection could be performed. An additional surgical procedure for the total facial paralysis was performed.

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