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Case report

Persistent stapedial artery in PHACE syndrome

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ABSTRACT

Introduction: PHACE syndrome is characterized by posterior fossa malformations, haemangioma, arterial anomalies, coarctation of the aorta, and eye abnormalities.
Case report: We present the case of a 6-year-old girl followed since birth for PHACE syndrome and left hemifacial haemangioma, who presented with left hearing loss. Computed tomography scan showed left persistent stapedial artery (PSA).
Discussion: Two types of arterial anomalies may be observed in PHACE syndrome: persistence of embryonic arteries and anomalies of cerebral arteries. PSA can be observed in the context of PHACE syndrome. Children with PHACE syndrome require regular audiometric follow-up to detect hearing loss and avoid its consequences on speech and language development.

1. Introduction

PHACE syndrome, first described by Frieden et al. in 1996, is a rare syndrome discovered at birth [1], comprising a combination of posterior fossa anomalies (P), haemangiomas (H), arterial anomalies (A), coarctation of the aorta (C) and eye abnormalities (E) [1].

Two types of arterial anomalies may be observed: persistence of embryonic arteries and anomalies of cerebral arteries [2]. No case of PHACE syndrome associated with persistent stapedial artery (PSA) has been previously reported in the literature.

We report a case of PSA discovered in the context of PHACE syndrome.

2. Case report

A 6-year-old child with a history of PHACE syndrome discovered at birth consulted for unilateral left hearing loss.

PHACE syndrome was diagnosed at birth in a context of tuberous segmental infantile haemangiomas of segments S1 (frontotemporal), S2 (maxillary) and S4 (frontonasal) of the left side of the face.

Brain magnetic resonance imaging (MRI) performed at birth revealed hypoplasia of the left cerebellar hemisphere with no contralateral or vermian lesion. The left vertebral artery arose directly from the aortic arch with no other anomalies of the rest of the circle of Willis. The child did not present any signs of heart disease or any eye abnormalities.

Clinical ENT examination revealed sequelae of the haemangioma of the cheek and left external ear (Fig. 1) with normal eardrums. Audiometry revealed conductive hearing loss of the left ear with a mean air-bone gap of 10 dB.

Unenhanced computed tomography (CT) of the temporal bones visualized a vascular channel arising from the left carotid artery ascending along the promontory of the tympanic cavity and passing between the two crura of the stapes, in contact with the anterior crus, before leaving the middle ear (Fig. 2). CT demonstrated absence of the left foramen spinosum, confirming the presence of a PSA giving rise to the left middle meningeal artery (MMA). A normal right foramen spinosum giving rise to the MMA was also observed.

In view of the child's minor hearing loss, management consisted of regular clinical and audiometric surveillance.

3. Discussion

PHACE syndrome is a rare craniofacial syndrome involving several consecutive segments of the neural crest. A dominant X-linked abnormality is suspected due to the female predominance of PHACE syndrome [3]. PHACE syndrome is usually diagnosed at birth in

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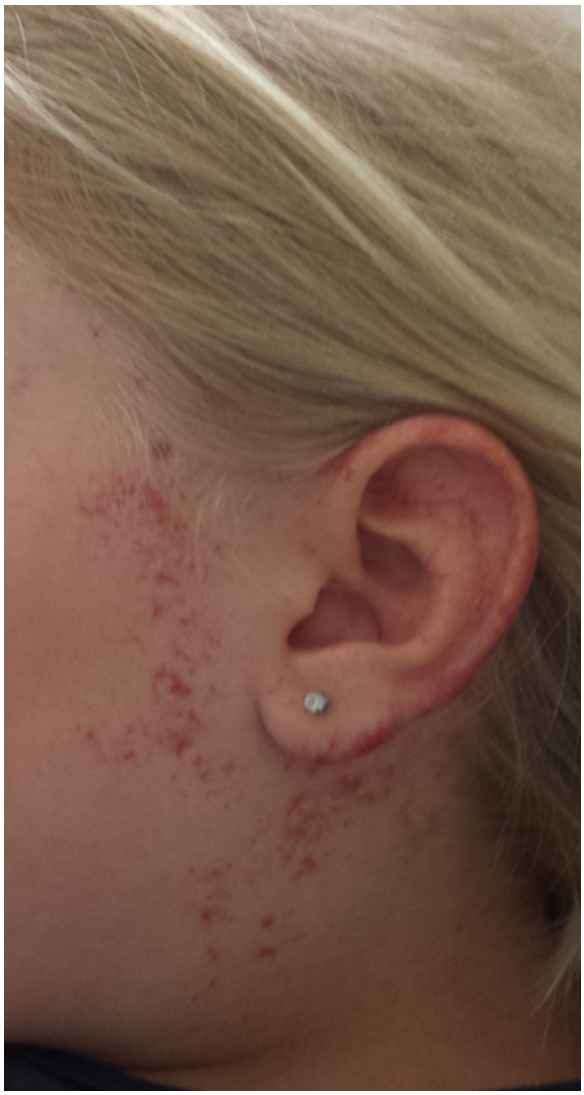

Fig. 1. Sequelae of the left hemifacial haemangioma involving the cheek and external ear.

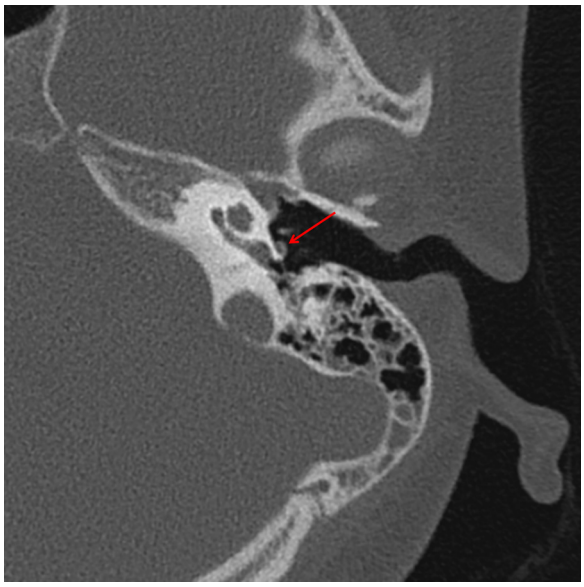

Fig. 2. Axial CT scan of the left temporal bone showing persistence of the left stapedial artery. Red arrow: persistent stapedial artery.

the presence of haemangiomas associated with posterior fossa lesions (cerebellar hypoplasia, arachnoid cyst, cortical dysgenesis or Dandy-Walker malformation) and arterial anomalies that are unilateral and ipsilateral to the haemangiomas in the great majority of cases [1,2].

Two types of arterial anomalies are observed:

- anomalies of cerebral arteries: dysplasia, stenosis, occlusion, hypoplasia or even agenesis of the carotid and vertebral and basilar arteries, aberrant origin or course, persistence of a trigeminal artery, saccular aneurysms;
- persistence of embryonic arteries, such as the proatlantal segmental artery, primary ophthalmic artery and primary hypoglossal artery [2].

PSA is a rare vascular anomaly, observed in about 0.48% of the population [4,5], corresponding to the persistence of an embryonic artery that normally involutes around the tenth week of pregnancy [5,6].

During embryonic development of the branchial system, six pairs of aortic arches are successively formed, connecting the aortic sac to the dorsal aortae. The second aortic arch gives rise to the hyoid artery (HA) arising from the dorsal aorta (future internal carotid artery). The stapedial artery arises from the HA at about 4-5 weeks of embryonic life. It then divides into a superior (supraorbital) branch, which subsequently becomes the MMA, and an inferior (infraorbital) branch, which subsequently becomes the infraorbital artery and the inferior alveolar artery. This inferior branch anastomoses with the ventral pharyngeal artery (future external carotid artery) arising from the aortic sac. The HA subsequently becomes the caroticotympanic artery, a branch of the internal carotid artery. Following involution of the stapedial artery, the MMA, arising from the external carotid artery, passes through the foramen spinosum. In the case of persistence of the stapedial artery, the MMA arises from this artery and therefore from the internal carotid system, leading to involution of the foramen spinosum [5,6].

A PSA is often an incidental finding during investigation of conductive hearing loss or pulsatile tinnitus and is responsible for stapes fixation secondary to pulsations of the artery on the crura of the stapes [5].

CT scan confirms the diagnosis by demonstrating a PSA associated with absence of the foramen spinosum, enlargement of the tympanic segment of the facial canal and sometimes prolapse of the internal carotid artery [4,5].

The case reported here did not present prolapse of the internal carotid artery, probably because PHACE syndrome is often associated with progressive stenosis of intracerebral arteries secondary to concentric proliferation of the vessel walls that gradually reduces the calibre of their lumen [2].

In the presence of symptomatic PSA with deafness or pulsatile tinnitus, several authors have described the use of electrocoagulation or laser coagulation with no complications, especially involving the facial nerve or hearing [4,5].

4. Conclusion

PHACE syndrome is a rare disease, which may be associated with unilateral persistence of embryonic arteries, including the PSA, ipsilateral to the haemangiomas. The diagnosis is usually based on temporal bone CT scan. These children require regular audiometric follow-up in order to consider coagulation of this artery if hearing loss becomes disabling and interferes with speech and language development.

Disclosure of interest

The authors declare no conflicts of interest related to this article.

References

[1] Frieden IJ, Reese V, Cohen D. PHACE syndrome. The association of posterior fossa brain malformations, hemangiomas, arterial anomalies, coarctation of the aorta and cardiac defects, and eye abnormalities. Arch Dermatol 1996;132(3):307–11.
[2] Rotter A, Samorano LP, Rivitti-Machado MC, Oliveira ZNP, Gontijo B. PHACE syndrome: clinical manifestations, diagnostic criteria, and management. An Bras Dermatol 2018;93(3):405–11.
[3] Assari R, Ziaee V, Moghimi S, Akbari MR, Mirmohammadsadeghi A. PHACE(S) syndrome: Report of a case with new ocular and systemic manifestations. J Curr Ophthalmol 2017;29(2):136–8.
[4] Breheret R, Bizon A, Tanguy JY, Laccourreye L. Persistent stapedial artery with otosclerosis. Ann Otolaryngol Chir Cervicofac 2009;126(5–6):259–63.
[5] Hitier M, Zhang M, Labrousse M, Barbier C, Patron V, Moreau S. Persistent stapedial arteries in human: from phylogeny to surgical consequences. Surg Radiol Anat 2013;35(10):883–91.
[6] Silbergleit R, Quint DJ, Mehta BA, Patel SC, Metes JJ, Noujaim SE. The persistent stapedial artery. AJNR Am J Neuroradiol 2000;21(3):572–7.