A complicated fourth branchial fistula

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A 5-year-old girl was admitted to our Medical Imaging Department for ultrasonography of a painful and non-erythematous cervical mass on the left side of the neck. She previously complained of pain and dysphagia for several days. There is no fever and no history of infection.

An axial gray-scale US (Fig. A) shows an irregular infiltrative hypo and hyper-echoic left cervical mass with adenopathies and deviation of the larynx. Colour Doppler US shows a moderate intratissuinal vascularization (Fig. B). Axial T1 and T2-weighted MR images (Fig. C) show an infiltrative parathyroidian mass of the left lateral neck (hypointense T1 and heterogeneous signal T2) with lateral displacement of the larynx, and a communication with the pyriform sinus. Barium esophagography (Fig. D) shows a left sinus tract from the apex of the pyriform sinus. These examinations confirm the diagnosis of a complicated fourth branchial fistula. Antibiotic therapy and chirurgical resection were performed and confirm the diagnosis.

Comment

Neck masses are a common finding in children and have a congenital, inflammatory, neoplastic or vascular origin. The fourth branchial arch contributes to the development of the larynx, which derives from the laryngotracheal groove formed during the 4th week of gestation. Anomalies of the branchial arch may appear as a cyst (extending from the apex of the pyriform sinus to the left thyroid lobe), a sinus (one opening-to-skin surface, external auditory canal, pharynx or hypopharynx), a fistula (connecting the skin to the lumen of the foregut) or an ectopic gland. Ninety percent of branchial anomalies arise from the second branchial arch. Anomalies arising from the fourth branchial arch are extremely rare and have been described in children and young adults. They are more common in females. The vast majority of these lesions occur on the left side. A sinus of the fourth branchial arch, following its embryologic origin, forms internally from the apex of the pyriform sinus, crosses the thyrohyoid membrane and the left thyroid lobe. It is sometimes difficult to distinguish a cyst of the third or the fourth branchial arch within the larynx. An internal fistula arising from the apex of the pyriform sinus pleads in favor of a fourth branchial pouch remnant.

Clinically, the lesion appears as a painless cervical mass in the posterior triangle area of the neck, anteromedial to the sternocleidomastoiden muscle. It can also manifest itself as a recurrent suppurative thyroiditis sometimes with dysphagia and stridor. Sonography is the imaging method of choice for evaluating congenital neck lesions in children. It shows a thin-walled echolucent cyst if not infected or a thick-walled hyperemic abcess in front of the thyroid. CT and MR are required to define the extent of the lesion. These examinations show either a thin-walled well-defined cyst without significant enhancement, or if infected, a thick-walled cyst with surrounding cellulitis. Barium esophagography or direct laryngoscopy may reveal an outpouching of the pyriform apex. A barium swallowing test can identify a sinus tract extending from the apex of the pyriform sinus to the lower anterior neck. Treatment consist of a chirurgical resection.

References