

TOLOSA-HUNT SYNDROME IN CHILDREN

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We report a case of Tolosa-Hunt syndrome in a 4-year-old girl. Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) revealed a left cavernous sinus involvement with internal carotid artery occlusion. Clinical signs included left ophthalmoplegia, convulsions and right hemiplegia. Etiopathogenesis of this syndrome is unclear but as in our case, response to corticosteroid therapy is spectacular and avoids unnecessary invasive diagnostic procedures. We underline the importance of cross-sectional imaging in the diagnostic approach. Several other diseases including tumors, vascular lesions and infections can have a similar clinical and/or imaging presentation. However, closely monitored evolution on imaging proves to be decisive in the establishment of final diagnosis of this syndrome.

Key-word: Cavernous sinus.

Tolosa-Hunt syndrome (TSH) is caused by proliferation of non specific inflammatory tissue in the cavernous sinus, superior orbit fissure or orbital apex. It manifests by painful ophthalmoplegia and paralysis of one or more oculomotor cranial nerves. TSH is characterized by rapid sustained response to corticosteroid therapy. In 1954, Tolosa reported the first clinical features and revealed a low-grade granulomatous inflammation in an autopsy of a patient with left ophthalmoplegia (1). In 1960, Hunt and al. reported the clinical criteria of Tolosa-Hunt syndrome (2).

The evolution of sectional imaging (CT and MRI) has encouraged the International Headache Society (IHS) to include the presence of abnormal inflammation tissue in cavernous sinus and orbital apex by MR in the diagnostic criteria (3). This implies that TSH is no longer diagnosed by elimination.

Case report

A 4-year-old girl presented with several episodes of convulsions. Clinically she had right-sided hemiparesis and contralateral ophthalmoplegia related to sixth nerve palsy. CT scan revealed a soft tissue mass in the left cavernous sinus extending to the skull base and enhancing strongly after contrast injection (Fig. 1). MRI also showed enlargement of the cavernous sinus occupied by abnormal tissue (hypointense on T1 and isointense on T2 compared to gray matter) and

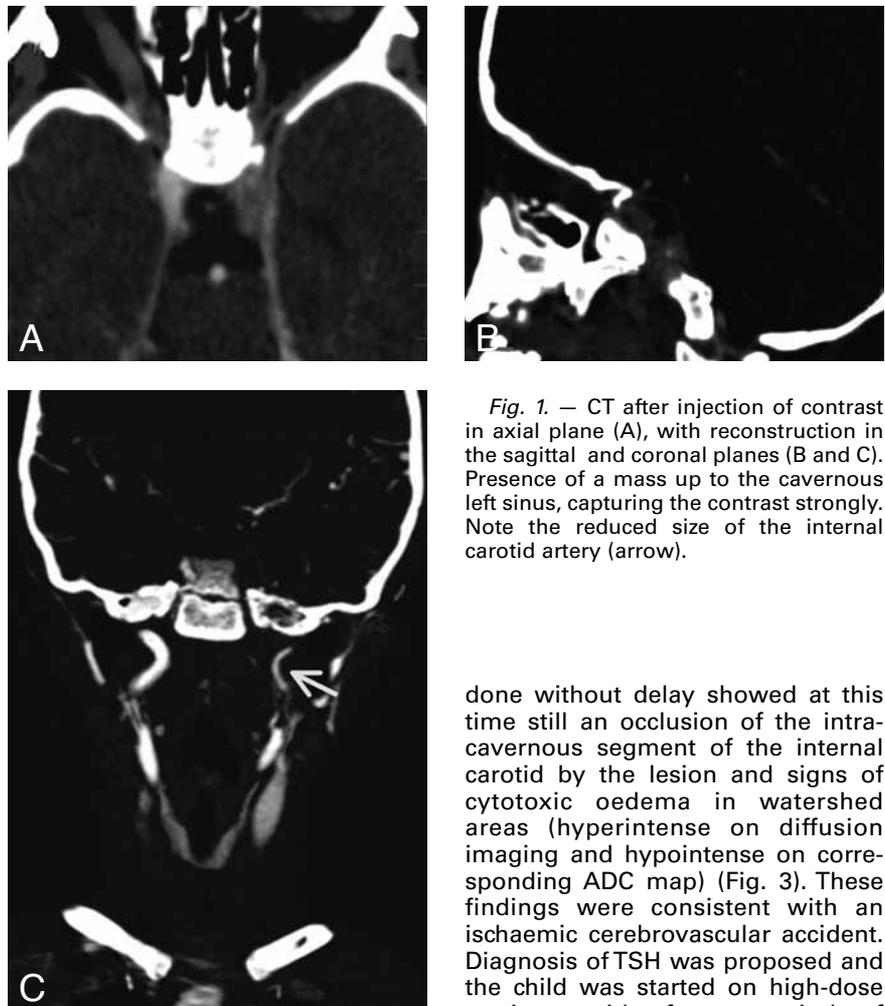


Fig. 1. — CT after injection of contrast in axial plane (A), with reconstruction in the sagittal and coronal planes (B and C). Presence of a mass up to the cavernous left sinus, capturing the contrast strongly. Note the reduced size of the internal carotid artery (arrow).

compression of the left internal carotid artery (Fig. 2). After a few days, the child experienced sudden and complete right hemiplegia. MRI

done without delay showed at this time still an occlusion of the intracavernous segment of the internal carotid by the lesion and signs of cytotoxic oedema in watershed areas (hyperintense on diffusion imaging and hypointense on corresponding ADC map) (Fig. 3). These findings were consistent with an ischaemic cerebrovascular accident. Diagnosis of TSH was proposed and the child was started on high-dose corticosteroids for a period of 2 months.

After that period, a control MRI showed few sequelae of the ischaemic lesion in the left hemisphere presenting as T2 hyperintensities in the white matter, compatible with gliosis. The soft tissue mass occupying the cavernous sinus had completely disappeared and the internal carotid artery looked patent and of normal size (Fig. 4). Clinically, the child had almost completely recovered from the motor deficits.

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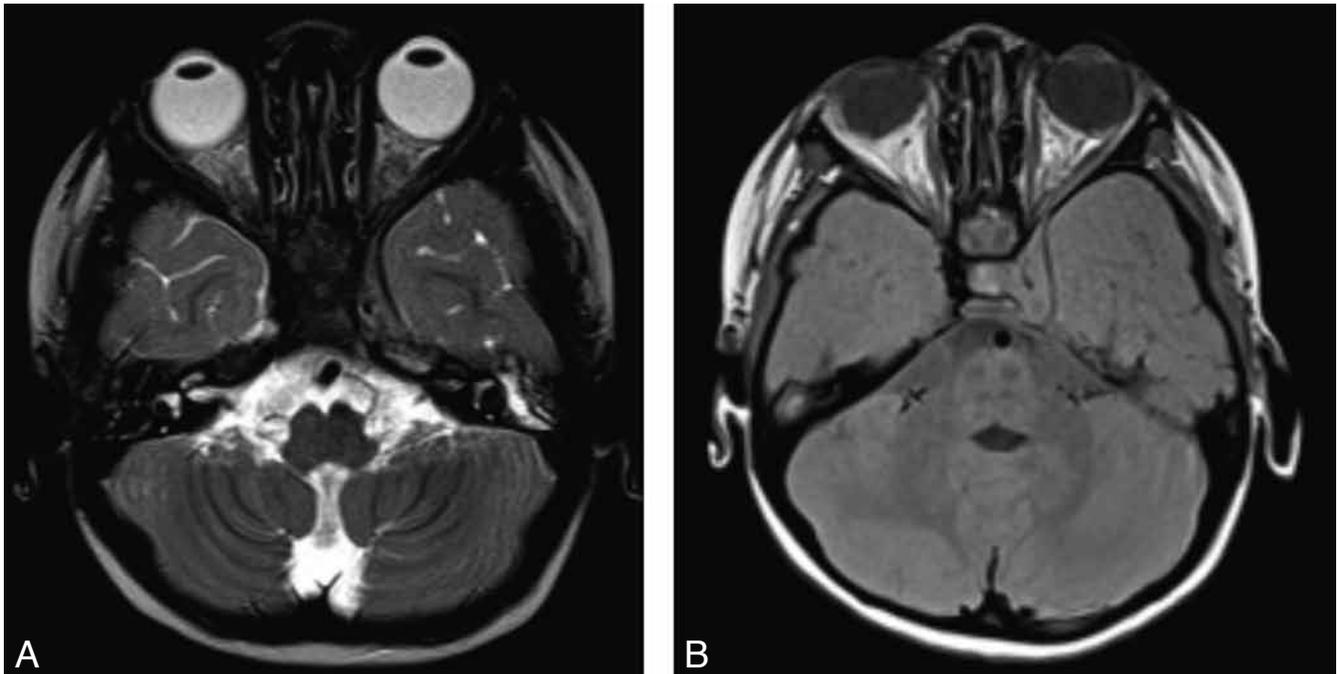


Fig. 2. — T2-weighted (A) and proton density (B). MRI showed a latero-sellar mass bulging the left cavernous sinus with occlusion of the internal carotid in its intracavernous portion.

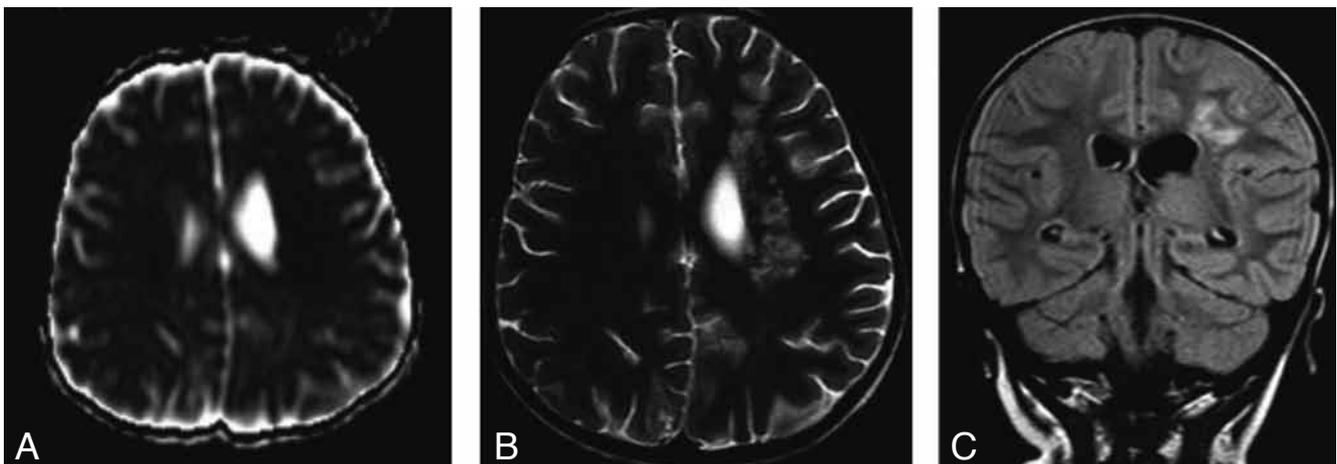


Fig. 3. — Diffuse involvement of the left deep white matter, hyperintense on diffusion with hypointensity on corresponding ADC map (A), T2 (B) and FLAIR (C) sequences in relation with ischemic lesion in the watershed area of the left internal carotid.

Discussion

TSH syndrome is defined as the presence of granulomatous and inflammatory soft tissue in the cavernous fossa, in the orbit apex or in the orbit itself (4). Its etiology remains unclear. As in our case, it is characterized by rapid response to corticosteroids and obviates the need for surgical exploration which can be hazardous due to small amount of pathological tissue and its relative inaccessibility. Clinical presentation which associates ophthalmic

plegia during more than 2 months and one or more cranial nerve palsy with rapid response to steroid treatment allows the diagnosis to be evoked (5), after having discarded other diseases with a similar presentation. These include aneurysm of the intracavernous segment of internal carotid artery, carotid-cavernous fistula, intra-cavernous meningioma, primitive lymphoma, tuberculosis and sarcoidosis (6).

CT or MR angiography can easily exclude an aneurysm of the carotid artery. If complicated, a carotid-cav-

ernous fistula can develop giving rise to exophthalmia and chemosis. In that case, MRI shows ophthalmic and cavernous veins dilatation (7).

Although rare in children, cavernous meningioma can give the same symptoms and presents the same features on MRI (hypointense on T1 and strong enhancement after injection) (8). Nevertheless, there is no response to steroid treatment.

Primitive lymphoma is rare and usually bilateral. It can particularly mimic THS as it also responds to steroid therapy (9). However, its

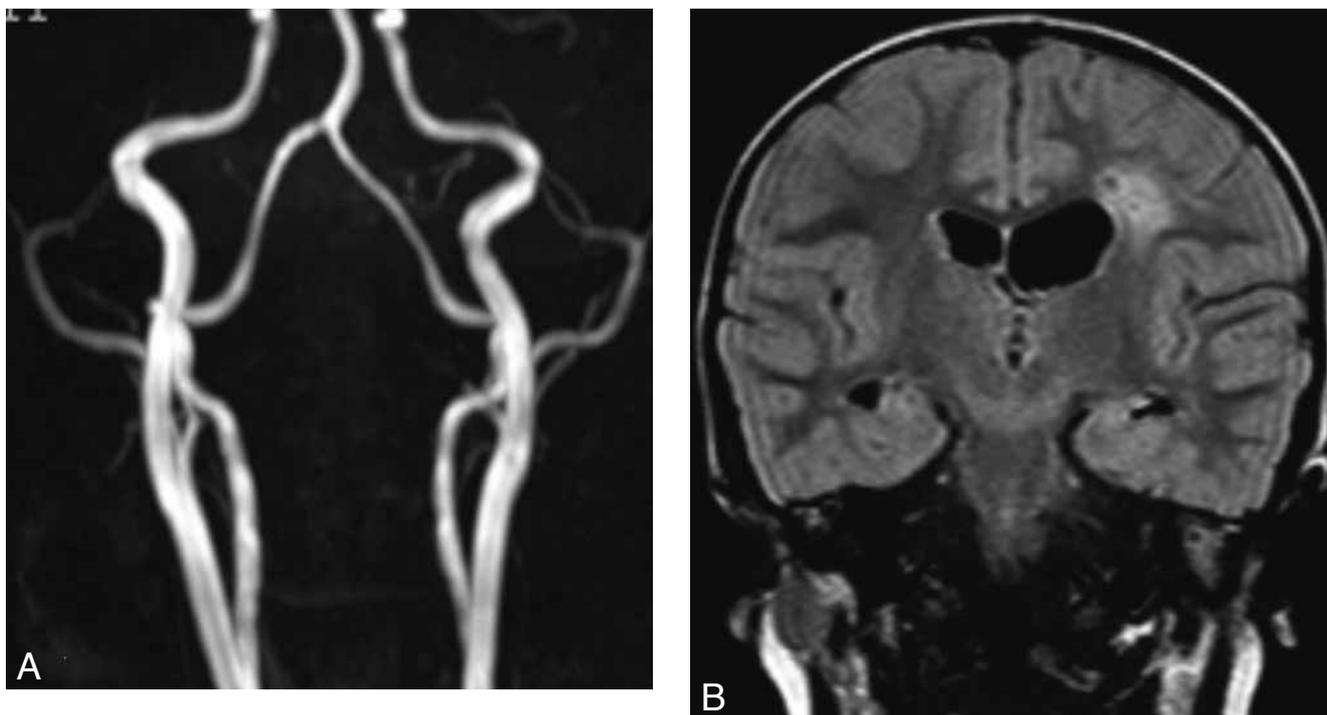


Fig. 4. — MR angiography (A) sequences demonstrating normal size of the left ICA. FLAIR sequence (B) showed hyperintense gliotic lesions.

diagnosis is usually made on the basis of other systemic signs. The same applies to tuberculosis and sarcoidosis.

Progress in cross-section imaging (CT and MRI) has been such that THS is no longer diagnosed by elimination alone. Characteristics of the lesions on MRI should meet precise criteria established by the IHS since 2004.

CT and MRI carried out in our patient have shown abnormal tissue (designated by Tolosa as being granulomatous) strongly enhancing after gadolinium injection and causing enlargement of the cavernous sinus with convex bulging of the external borders of the dura-mater. These correspond to the 3 primary criteria of THS on MRI (10). Low intensity on T1 imaging and strong contrast enhancement are in agreement with other case published in the literature (11).

As in the first case published by Tolosa in 1954, we found a stenosis of the intra-cavernous segment of one internal carotid artery in our patient. Potentially damaging cerebral lesion stresses on the gravity of this syndrome and on the importance of an early diagnosis. We also

highlight the use of an early and aggressive steroid therapy (5) which, in our case, almost completely resolved the lesions and the symptoms.

Conclusions

TSH is a rarely reported affection in children. Its consequences can be damaging especially by the occurrence of cerebral complications. Early diagnosis can be established by associating the clinical setting and the use of cross-sectional imaging. Steroid therapy can hence be started without delay, with spectacular results.

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