Lateral sinus thrombosis as a complication of acute mastoiditis

Trombosi del seno laterale come complicanza di mastoidite acuta

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Summary

Lateral sinus thrombosis is a rare complication of middle ear diseases: in children, it is usually related to acute otitis media, but it is also found in adults with chronic otitis. It was more frequent in the pre-antibiotic era and mortality was high. The Authors present a paediatric case of lateral sinus thrombosis in which they describe the clinical approach and related literature.

Key words: Acute otitis media • Complications • Lateral sinus thrombosis • Diagnosis • Angio-MR • Anticoagulant therapy

RIASSUNTO

La trombosi del seno laterale è una complicanza rara delle patologie dell’orecchio medio: in età pediatrica si correla solitamente ad una quadro di otite media acuta, mentre nell’adulto può associarsi anche ad un’ottite media cronica. La trombosi del seno laterale era di più frequente riscontro in era pre-antibiotica, con mortalità elevata. Gli Autori descrivono un caso di trombosi del seno laterale in età pediatrica, presentando una revisione della letteratura.

Parole chiave: Otitis media acuta • Complicazioni • Trombosi del seno laterale • Diagnosi • Angio-RM • Terapia anti-coagulante

Introduction

Lateral Sinus Thrombosis (LST) was considered a frequent complication of middle ear infection at the beginning of the last century and mortality reached up to 100% in untreated cases. Over the last fifty years, its incidence has greatly decreased due to the widespread availability of antibiotic drugs and mortality has dropped below 10%. Morbidity, related to LST, is approximately 30% and is associated with septic cardiomypathy, acute respiratory distress syndrome, anacusis and seizures. Antibiotic resistance has now been recognized as the main factor of the increasing incidence of LST as a complication of acute and chronic otitis. Especially in young adults, LST is now more often seen in association with a generalized hypercoagulable state, inherited or acquired.

The Authors present a paediatric case of LST in which they describe the clinical approach and the related literature review.

Case report

BB, a 4-year-old girl, was admitted to the ENT Department in November 2001, with a history of a left acute otitis media which had built up over the past 4 days and with a temperature fluctuating between 38.5 °C-40 °C. On examination, a purulent otorrhea, as well as a significant left post-auricular swelling, were present. Computed tomography (CT) scan of the brain and temporal bones without contrast was immediately performed revealing diffuse clouding of left tympanic and mastoid cavities, without signs of coalescent mastoiditis or bone erosion. Laboratory investigations showed an inflammatory syndrome (WBC 10400/µl, PCR 17.00 g/dl, VES 112 mm). During paediatric hospitalization, the child received antibiotic therapy intravenously (iv) (Ceftriaxone 1 g/day). An auricular specimen, collected for microbiologic test, resulted negative. Over the following days, the local and general conditions of the patient improved and after 6 days the patient was discharged with antibiotic therapy (amoxicillin and Clavulanate 1 g perorally/day). Upon examination, 2 days later, swelling in the left retroauricular area and a fever were observed (37.5°); microtoscopy indicated persistence of the acute involvement of the middle ear. The patient was hospitalized. Myringotomy and enlarged mastoidectomy were performed; there was evidence of partial erosion of the bone covering the lateral sigmoid sinus area. A retroauricular drainage was inserted and removed after 4 days. The intra-operatory microbiological sample was negative. Antibiotic treatment iv was changed (Ceftazidime 600 mg/day, Amikacin 250 mg twice a day, Teicoplanin 180 mg/day). After surgery, the clinical status suddenly improved.
and no fever was present. A post-operative (CT) scan of the brain and temporal bones, without contrast, was performed 5 days after surgery, to clearly define the extension of the bone erosion and to exclude cerebral involvement. Imaging showed a wide erosion involving the left mastoid and sinus plate with no evidence of cerebral lesions. Magnetic resonance imaging (MRI) revealed thrombosis in the left lateral and transverse sinuses and meningeal inflammation, with thickening and contrast enhancement (Figs. 1A, 2A). Magnetic resonance angiography (MRA) confirmed the lack of the normal high flow in the left lateral and transverse sinuses (Fig. 3A). The study of the hypercoagulability status was normal. The patient received low molecular weight heparin (0.3 ml per day). After 20 days, MRI showed the persistence of thrombosis in the lateral and transverse sinuses. Anticoagulant therapy was then administered for the following 5 months (low molecular weight heparin 0.3 ml per day), in accordance with haematologic consultation. After a further 6 and 12 months, MRI and MRA were performed and showed partial recanalization of the involved venous sinuses and normalization of the meninges (Figs. 1B, 2B, 3B).

Fig. 1A. T1 weighted MR axial images after contrast administration. MR performed a few days after surgery displays constrast enhancement of left lateral sinus, that has lost normal slow flow signal.

Fig. 1B. T1 weighted MR axial images after contrast administration. MR performed one year later displays normal flow-related signal in central part of left lateral sinus.

Fig. 2A. T1 weighted MR coronal images after contrast administration. MR performed a few days after surgery displays meningeal thickening and contrast enhancement in left middle cranial fossa, without lesions involving temporal lobe; jugular vein bulb is enhanced after contrast.

Fig. 2B. T1 weighted MR coronal images after contrast administration. MR performed displays no meningeal enhancement in middle cranial fossa and normal flow related signal in jugular bulb.

Fig. 3A. MRA supero-inferior view. A few days after surgery, no flow is displayed in left transverse and lateral sinuses and internal jugular vein bulb.
Discussion

Lateral sinus thrombosis is usually associated with acute or chronic middle ear diseases. As far as concerns pathogenesis, it has been proposed that the propagation of infection from the small venules of the mastoid into the sigmoid sinus could result from direct spreading of the inflammatory process through a coalescent or cholesteatomatous bone erosion, causing the formation of a perisinus abscess. Subsequently, adherence of fibrin, blood cells and platelets can produce a mural thrombus. The persistence of the inflammation and the increase in thrombus volume might result in an oblitative LST. In the early course of acute otitis media, LST can be caused also by an osteostrombophlebitis phenomenon; if this is the case, the sigmoid bony sinus plate will be intact at the time of surgical exploration.

Hypercoagulable states, inherited (antithrombin III, protein C or protein S deficiency, factor V Leiden mutation or prothrombin gene 20210A mutation) or acquired (neoplasms, trauma, autoimmune disorders, neurosurgical interventions, myeloproliferative disease, anti-phospholipid syndrome, pregnancy or use of oral contraceptives) are considered as risk factors in LST. Thus, once LST has been diagnosed, investigations to identify all possible haematological predisposing risk factors are necessary.

Several microbiological agents can be isolated and cultured in LST cases; Sym et al. reported anaerobic organisms in 100% and Proteus Spp in 66% of patients with LST secondary to chronic otitis media. Seven et al. described Proteus Mirabilis followed by Pseudomonas Aeruginosa and Bacteroides Fragilis as the most common microorganisms detected, while in one third of patients no germs could be found. Also B-haemolytic Streptococcus and Staphylococcus Aureus have been reported as causative agents.

At least two clinical pictures, related to LST, are described: the septic form and the aseptic form. The first is associated with clear signs of otomastoiditis and rarely complicated by cerebral abscess. The aseptic form is often associated with endocranic hypertension and possible ocular signs. The case described is a typical occurrence of the septic form.

The most common manifestation in patients with LST is fever, which is usually sustained or occurs in a spiking pattern. Other symptoms are otorrhea, post-aурicular oedema, otalgia, headache, nausea, vomiting and meningal signs. Papilloedema and the Griesinger sign, consisting of oedema and tenderness over the mastoid process (due to the septic thrombosis of the mastoid emissary veins) are often present. LST can be also associated with sixth and seventh nerve palsy, hydrocephalus or signs of thrombus propagation, that can also cause, proximally, jugular bulb and, distally, occlusion of other dural sinuses and then neurological symptoms due to involvement of the lower cranial nerves (IX, X, XI pairs).

When clinical suspicion of intracranial complications, during middle ear infection, is present, imaging is considered essential to formulate diagnosis and planning management. Contrast-enhanced CT imaging of the brain and temporal bones displays the sinus plate erosion and may show the “delta sign” (central non-enhancing clot surrounded by enhancing dural sinus wall), indicating sinus thrombosis, and other intra-cranial complications, such as brain abscess and empyema. In the diagnosis of LST, MRI is considered to have a higher resolution than CT on account of its ability to show low or absent flow in the venous sinuses, clot formation and the presence of inflammation in the brain and meninges. Using MRI, the presence of a thrombus appears as an increased signal intensity in T1 and T2 images. MRI is useful in confirming the diagnosis of thrombosis in doubtful cases on MRI.

Treatment of LST has recently become more conservative. Most Authors agree that, in children, surgical treatment of septic LST combined with antibiotic therapy i.v. is standard care, today. Mastoidectomy is performed in patients with AOM, while radical or radical modified mastoidectomy is recommended in cases of cholesteatoma, depending on the extension of the disease. A combined neuro- and oto-surgical approach has been suggested, instead, to drain the purulent discharge and to remove the thrombus.

Internal jugular vein ligation using a cervical approach to prevent thrombus propagation, routinely performed in the pre-antibiotic era, is now controversial. Today, current indications to this treatment are persistent septicaemia even after mastoidectomy, or septic pulmonary or extrapulmonary embolization. Since the risk of embolization is low, jugular vein ligation is rarely necessary. Recently, anti-coagulant treatment has been proposed to prevent complications associated with thrombus persistence or its possible propagation. If anti-coagulation use is mandatory in all patients with a hypercoagulable state, inherited or acquired, the role of anticoagulants in otogenic LST cases is still not clear, be Bradley et al. pointed out that the risk of complications related to LST (such as embolization and persistent sepsis) should be balanced with the possible complications associated with anti-coagulation, especially within the paediatric population, as major risks of anti-coagulation treatments include thrombocytopenia, drug interactions, bleeding, haemorrhagic skin necrosis and bleeding. Nevertheless, if anti-coagulant therapy is necessary, then the type of drug used, the entire duration, the dosage, as well as the specific patient characteristics, must be taken into consideration.

In the case presented here, mastoidectomy associated with anti-coagulant treatment, represented the therapeutic approach correlating with extension of disease and local complications.

No other treatment was necessary.
Lateral sinus thrombosis

Conclusions
LST in children is considered an unusual complication of otitis media requiring accurate recognition and treatment. Imaging is essential to formulate diagnosis and for planning strategy. Management of LST is controversial, but usually consists of conservative surgical treatment combined with antibiotic therapy iv. Recently, anti-coagulant treatment has also been proposed.

References

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