Pseudomastoiditis in Lateral Sinus Thrombosis: A Rare Presentation with Review of Literature

Ramadass Thota · Nithya Narayanan ·
Dhanraj Mathuram

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Abstract Lateral sinus thrombosis due to mastoiditis, is now a relic of the past. Gone are the days when septic lateral sinus thrombosis was a common complication of unsafe suppurative otitis media. In the present era, the neurologists more commonly see cortical venous thrombosis with lateral sinus thrombosis. This entity has been termed as non-septic lateral sinus thrombosis in literature. The incidence is found to be more in females due to the use of oral contraceptives (Rosen and Scher in *Laryngoscope* 107(5):680–683, 1). In this manuscript, we report a series of three cases of non-septic lateral sinus thrombosis with mastoiditis, seen in a span of 1 month, which is uncommon. All the patients presented to the neurologist with intractable headache and lateral sinus thrombosis with mastoiditis, was detected by magnetic resonance imaging and magnetic resonance venogram. All the three patients were males and had involvement of the right mastoid in magnetic resonance imaging pictures. We have reviewed the literature and discussed about the etiopathogenesis, diagnostic criteria and management.

Keywords Lateral sinus thrombosis · Mastoiditis · Nonseptic lateral sinus thrombosis · Cortical venous thrombosis · Magnetic resonance venogram

Introduction

In the preantibiotic era, lateral sinus thrombosis (LST), was a common complication of suppurative ear disease with

mastoiditis and sinus infections. LST is a complication of extensive cholesteatoma eroding the sinus plate, resulting in thrombophlebitis. Lateral sinus comprises of sigmoid sinus and transverse sinus. Intracranial veins and venous sinuses do not have valves and blood within them can flow in either direction, depending on haemodynamics [1]. A mural thrombus formation can block the sigmoid sinus and can spread proximally to the transverse sinus, torcular herophili, superior sagittal sinus, superior petrosal sinus and the cavernous sinus. It can also spread distally to involve the jugular bulb, internal jugular vein and subclavian vein [2]. The nagging headache and pain in the mastoid region radiating to the neck, with constitutional symptoms like high fever, with chills and rigor (Pickett Fence pattern) and intracranial complications, heralds the disease process in septic LST [3].

In the recent times, such complications are sparsely seen because the attending physicians treat the ear infections extensively with antibiotics and the disease may be dormant. Intractable headache and neurological complications prompt the patients to consult the neurophysicians. In this presentation we report three cases of cortical venous thrombosis (CVT) with LST with mastoiditis, detected by magnetic resonance imaging (MRI) by the neurologist and referred to us to rule out septic causes in ENT. All the cases were thoroughly examined and there was no evidence of sepsis in ears and sinuses. The mastoiditis like radiological picture had occurred due to venous congestion and subsequently cleared up with conservative treatment. The mastoiditis in CVT is a spurious one, as the ear is clinically normal. We reviewed the literature through Internet and found that nonseptic lateral sinus thrombosis (NSLST) cases are on the increase and a radiological finding of mastoiditis without clinical symptoms as seen in our patients, is a rare phenomenon.

R. Thota (✉) · N. Narayanan · D. Mathuram
Apollo Hospitals, Chennai 600 006, India
e-mail: ramadassthota@yahoo.com

Case Reports

Case 1

A 25 year old gentleman, presented with bitemporal headache of one-week duration. Headache started with fever, which subsided on its own. Patient had ear pain on both sides with diplopia. Examination revealed a septal deviation to right with normal tympanic membrane, absence of mastoid tenderness and normal hearing. Neurological examination showed right lateral rectus palsy and other cranial nerves were found to be normal. Ophthalmologist's opinion was sought and fundoscopy showed papilloedema on both sides. MRI and magnetic resonance venogram (MRV) of brain showed right sigmoid sinus and transverse sinus thrombosis with right mastoiditis. X-ray mastoids showed a partly sclerosed right mastoid and a cellular left mastoid. Laboratory investigations including coagulation profile were normal. In view of the MRI report of mastoiditis, a mastoidectomy was planned. The patient was not willing for surgery and hence the neurologist treated him as an outpatient, with oral anticoagulants. The patient improved symptomatically and a follow up

computerized tomographic (CT) scan of temporal bones was found normal.

Case 2

A 28-year-old gentleman presented with headache of 4 days duration with nausea. He had transient slurring of speech and altered sensorium for 1 day. There was no vomiting, fever, fits or ear discharge. Clinically he was conscious, oriented, flexor plantar response was elicited on right side and extensor on left side. Mild neck stiffness was present. Fundus examination revealed bilateral papilloedema.

MRI of brain with MRV revealed right mastoiditis (Fig. 1) with thrombosis of superior sagittal sinus, both lateral and sigmoid sinus and partial thrombosis of straight sinus, with diffuse cerebral oedema. Laboratory tests including coagulation profile were normal. Clinically ENT examination was found to be normal. Audiogram showed a bilateral high frequency sensorineural dip at 4 kHz.

He was admitted in ICU by the neurologist and treated with Heparin 5000 units subcutaneously twice a day, along with sodium and potassium supplements, acenocoumarol, acetazolamide and acyclovir. Mastoid exploration was

Fig. 1 T2 weighted MR image of brain showing hyperintense signal in the right mastoid air cells

planned at a later date as patient was on anticoagulants. Subsequently he was discharged and patient came to us for follow up after a month. His symptoms had abated by then. A CT scan of temporal bones showed a normal mastoid bone on both sides.

Case 3

A 37-year-old gentleman presented in emergency with two attacks of right focal seizures, vomiting, loose stools and headache in occipital region. On the day of admission he suddenly developed restlessness, confusion and right focal jerky movements. There was no history of fever, visual disturbance or ear discharge. On admission he was conscious, obeying commands but was confused. He had right upper motor neuron facial weakness and slurred speech. No neck stiffness was noted. Fundus examination revealed bilateral papilledema. ENT examination did not reveal any clinical evidence of ear disease and mastoid tenderness. His audiogram was normal.

MRI of brain with MRV (Fig. 2) revealed acute to subacute stage of infarct in posterior parietal region with right mastoiditis, thrombosis of superior sagittal sinus and

straight sinus extending up to lateral and sigmoid sinus, with diffuse cerebral oedema and a corpus callosum lipoma. All laboratory tests were normal, except a blood sugar of 182 mg% and HBA₁C was 8.2%. His coagulation profile was normal. A diabetologist was consulted for glycemic control. He was treated with Inj Heparin 5000 units subcutaneously twice a day, Tab Phenytoin sodium, Inj Mannitol, Tab Acetazolamide and switched over to oral anticoagulant—Acenocoumarol (3 mg) once a day, during discharge. During follow up, by 3 months he had recovered well and had mild dysphasia only. Repeat MRI of brain done after 6 months showed a normal right mastoid.

Discussion

We have described three cases of NSLST with mastoiditis, referred to us for evaluation of septic focus by the neurologist. Nonseptic lateral sinus thrombosis (NSLST) is an entity, which differs from septic lateral sinus thrombosis (LST) in that it is not associated with any ear or sinus infections. Rosen and Scher had reported three cases of NSLST, all in female patients occurring on the left side due

Fig. 2 MR intracranial venogram showing thrombosis of superior sagittal sinus, both lateral and sigmoid sinus with multiple venous collaterals

to the usage of oral contraceptives [4, 5]. It has also been found to be associated with many other conditions namely thrombocytosis, sickle cell anemia, altered cerebral hemodynamic states, dehydration from vomiting, thyroid disease, immunologic abnormalities including the presence of circulating antiphospholipid antibodies etc. [6, 7]. Haemodynamic disturbances caused due to circulating infective emboli can cause disturbance to venous drainage from inside the cranial cavity [8]. Mostly young women in pregnancy or puerperium are affected. The risk of developing NSLST is also high in women using oral contraceptives and the risk is compounded if they are smokers. Autopsy studies have demonstrated endothelial proliferations in dural sinus walls adjacent to the thrombus, induced by the estrogen component in oral contraceptives. This can cause enhanced deposition of fibrin in the endothelial lining predisposing to thrombus formation.

The usual mode of presentation in these patients with NSLST is intense headaches—generalized or localized to the temporoparietal area, corresponding to the side of the thrombus and ear pain [9]. With extension of the thrombus in the dural sinuses, the symptoms of raised intracranial pressure may occur. The typical spiky fever due to release of emboli (Pickett Fence pattern) in septic LST is absent in these patients. Most patients are in the age group of 40–50 years. Though the ear was found normal clinically, the MRI picture showed mastoiditis in all three patients. The patients had no mastoid tenderness. The finding of mastoiditis radiologically is essentially due to mucosal edema in mastoid air cells. The venous channels normally pass through a thin bony plate separating the mucoperiosteum of mastoid air cells from the sigmoid dural sinus wall. The possible reason described for the mucosal edema is impaired venous drainage from the mastoid cells into the lateral sinus due to the thrombus formation, which blocks these channels. This leads to vascular congestion, interstitial edema and transudation of fluid into air spaces [8]. Conservative treatment has been recommended for NSLST with asymptomatic mastoiditis.

In our study, the three patients had NSLST with pseudomastoiditis affecting the right side only. All the patients were investigated thoroughly but the exact predisposing cause of the sinus thrombosis with mastoiditis, in these patients could not be established. Our presumption is that an altered haemodynamic state may be responsible. In these patients, the MRI report showed mastoiditis on the right side without any clinical signs and symptoms. Nevertheless, mastoid exploration was planned but postponed because they were on anticoagulants. Eventually they became better symptomatically with conservative treatment and follow up CT scan of the mastoid bone was normal. In view of these bizarre findings which mimicked mastoiditis, it is appropriate to name this condition as

Pseudomastoiditis which is a rare phenomenon described in literature.

MR venogram is the gold standard for diagnosis of NSLST [10]. T2 weighted MRI is sensitive to mucosal changes and accumulation of fluid in the mastoid air cells. T2 weighted MRI signals in mastoid air spaces are seen as a trabecular pattern consistent with mucosal thickening in the air cells [8, 11]. Contrast CT also has good sensitivity to pick up dural sinus thrombosis. Review of literature shows that the presence of a radiological finding mimicking mastoiditis in a patient with cortical venous thrombosis, with a normal ear can be treated conservatively with anticoagulants [12]. In our series, surgical treatment was not necessary and the patients improved considerably, after treatment with anticoagulants. The interesting feature is that all these three patients were seen in a span of 1 month whereas such cases are rare to see in routine clinical practice and the role of MRI is remarkable. Repeat MRI scans of patients with NSLST during follow up, is necessary to know the condition of the mastoid. In majority of the cases NSLST is generally treated with anticoagulants alone and invasive surgery should be avoided unless indicated [13].

Conclusion

Nonseptic lateral sinus thrombosis with radiological features of mastoiditis and without any clinical evidence of mastoiditis is a rare phenomenon encountered in clinical practice. It presents with intractable hemicranial headache and is diagnosed by MRI and MRV. The otolaryngologist is consulted to rule out ear and sinus infection. MRI is the only tool for diagnosis of this disease, besides the clinical presentation of the disease. Treatment of NSLST is necessary to prevent propagation of thrombus to avoid thromboembolism. A conservative approach for treatment is adopted, with anticoagulants. The important message to take home is that the mastoid findings in NSLST are secondary to venous congestion; hence it is a pseudomastoiditis, which does not warrant surgical treatment.

Key Messages

- Non-specific lateral sinus thrombosis with mastoiditis is a rare entity, on the increase.
- Non-specific headache, ear pain, neck pain prompt the neurologist for imaging studies.
- Pseudomastoiditis occurs due to blockage of draining mastoid veins.
- Etiologic factors include hypercoagulable states, altered hemodynamic states etc.

- Non-septic lateral sinus thrombosis is symptomatically different from septic lateral sinus thrombosis.
- Treatment is always conservative with anticoagulants.

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