Two cases of dural sinus thrombosis and review of the literature

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Abstract: Dural sinus thrombosis (DST) may occur due to infection, surgery, hypercoagulability or compression of the sinuses by tumors. DST in the cases of intracranial hematoma may also occur after head injury such that depressed skull fractures or skull fractures that cross the sinuses obstruct the blood flow in the sinus.

DST is rare with wide range of non-specific signs and symptoms, ranging in severity from mild headache to coma and death. We present two cases of DST in this report and review the literature for this condition which has high risk of morbidity and mortality. The first case was a 16 years old boy hospitalized because of history of headache, nausea and repeated vomiting for two day after a motorcycle traffic accident who died on the third day of his hospitalization. The second case was an 18 years old girl who had a history of upper respiratory infection and diarrhea for four days and died several hours after her admission to emergency service. In both cases, DST was diagnosed at medicolegal autopsy.

Key Words: dural sinus thrombosis, head injury, septic thrombosis.

Dural sinus thrombosis (DST) was diagnosed post-mortem by Ribes in 1825 for the first time [1]. Thrombosis of the dural sinuses and cerebral veins was until recently considered a rare, often fatal disease, related to the puerperium and to the infection of the central nervous system, sinuses, and mastoids [2, 3].

Diagnosis of DST was difficult until the end of the last century, because an invasive technique such as autopsy or invasive X-ray angiography was required. As a result of recent advances in diagnostic tools such as Computerized Tomography (CT) and Magnetic Resonance Imaging (MRI), DST is more frequently diagnosed [1].

The estimated annual incidence of DST is 1.5 to 4 cases per million in adults and 6.7% per million in children [1, 4]. The prevalence is only 1% in consecutive autopsies [3]. There are many etiological risk factors associated with DST, which include hypercoagulable states, oral contraceptive usage, pregnancy, puerperium, infection, compression of the sinuses by tumors, arteriovenous malformations, surgery and mechanical causes such as cranial trauma [4, 5, 6].

In head trauma, cerebral sinus occlusion is induced not only by thrombosis but also by direct compression caused by a depressed skull fracture or epidural hematoma and skull fractures that cross the sinus that obstruct the blood flow [7].

Severe dehydration, inflammatory bowel diseases, connective tissue disorders, sarcoidosis, nephrotic syndrome, hormonal replacement therapy, steroids, oncologic treatments, lumbar puncture and parenteral injections may also lead DST [1, 5, 8]. Similarly DST presents with a wide spectrum of symptoms and signs which might be difficult to differentiate from many other disorders [5, 6, 8].

Clinical symptoms and signs depend on the location and the number of occluded sinuses and veins. In addition, existence of parenchymal lesions, age of the patient as well as the interval from onset to presentation also determines the signs and symptoms [3]. The most common symptoms and signs are headache, seizures, focal neurological deficits, altered consciousness, and papilloedema, which can present in isolation or in association with other symptoms [3, 8].

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Because of its nonspecific clinical manifestations and radiological findings, diagnosis of DST is still a problem. Delay in diagnosis may lead to venous congestion, venous infarction, and even death [9]. Therefore, early and accurate diagnosis is necessary to save patient’s life.

In the last 7 years, there were two cases of dural thrombosis among the 3493 medicolegal autopsies performed by Morgue Department of Antalya Branch of the Forensic Medicine Council. We present these two rare cases of DST diagnosed at medicolegal autopsy and we also review the literature for this condition in this report which was previously presented as a poster in the 22nd IALM Congress and published in abstract form [10].

CASE 1

A 16 years old boy hospitalized because of history of headache, nausea and repeated vomiting for two days after a motorcycle traffic accident who died at third day of his hospitalization. In his first admittance, he had a history of somnolence, temporary memory loss and he had bruises and abrasions on right and left upper rear side of his scalp and ulnar fracture. He had been referred to a tertiary hospital. According to his medical records, his neurological examination as well as non-contrast CT did not reveal any abnormalities. He was discharged asymptomatic after observation for 9-10 hours. After three days of the accident, he had readmitted to the hospital with headache and decreased level of consciousness. After physical examination, which revealed rhinorrhea and the signs of meningeal irritation, LP was performed. There were plenty of leucocytes on CSF. After his initial examination his condition deteriorated and he became unconscious. He was intubated and transferred to the intensive care unit. Hyperdense collection on the left sphenoid sinus was found while cerebral and cranial structures were reported to be normal at CT. Gram positive cocci were detected and pneumococci were isolated in the CSF culture. Hypernatremia developed while he was taking antibiotics and anti૚oedema treatment. He was accepted as brain death on his third day of his second hospitalization.

Medico legal autopsy was performed. There were ecchymosis and abrasions on the rear side of both temporoparietal area of his scalp. Epidural hematoma was present on the right anterior part of middle fossa of the basis cranium. Dura was dull and yellowish on this area. There was a fracture line from orbital plate of frontal bone to small wing of sphenoid at the right anterior fossa.

Bone fragment (0.3x1 cm) which had been broken off from small wing of sphenoid was found in the hematoma. Fracture line was extended to medial part of giant wing of sphenoid. All dural sinuses, especially cavernous sinus, right lateral sinus and sagittal sinus were occluded with the thrombus. Histopathological examination confirmed the widespread thrombosis and menengitis. In addition to these, septic thrombosis was determined especially in the cavernous sinus. Toxicological tests didn’t reveal any toxic or abused agent.

CASE 2

According to medical records, an 18 years old girl who had a history of upper respiratory infection, weakness and diarrhea for four days, was examined and treated with ornidazole, rabeprazole, and metamizole sodium. Two days after her first examination, she had readmitted with sore throat, headache and fever. She was diagnosed with acute pharyngitis and ibuprofen and pseudoephedrine were prescribed. The following day she was readmitted to the hospital. The general condition worsened during her admittance and she became unconscious. She was resuscitated and during intubation, fresh blood came out of the endotracheal tube. Chest radiography and thorax CT showed interstitial pulmonary infiltration. After 3 hours of admittance, she went into a cardiac arrest and there was a massive bleeding from the endotracheal tube and she died.

The laboratory test results were as follows: WBC 73.85 (N:4.8-10.5), NEUT 47.04 (N:2.2-4.8), LYMP 21.27 (N:1.3-2.9), MONO 4.8 (N:0.3-0.8), BASO 0.22 (N:0.0-0.1), RBC 2.95 (N:4.7-6.1), Hemoglobin 6.3 g/dL (N:14-18), Hematocrit % 20.8 (N:42-52), PLT 171 (N:150-300), Sedimentation % 21 (N:0-20), INR 1.29 (N:1-1.4), fasting blood glucose 265.8 mg/dL (N:70-110), Direct bilirubin 0.53mg/dL (N:0.1-0.4), AST 71,86 U/L (N: 0-32), ALT 59.54 U/L (N<35), LDH 699 U/L (N<248), Calcium 8.33 mg/dL (N:8.8-10.5), CRP 38.7mg/dL (N: 0-5) and creatinine, uric acid, total bilirubin, GGT and urea level and percentile values of Neutrophil, lymphocyte, monocyte, basophil were normal.

There was no physical sign of trauma on her body.
at the medico legal autopsy. Both lungs were heavy (right 1018 g, left 882 g) with bleeding and firm. Subarachnoidal hematoma in 0.5 cm diameter was found on the surface of right parietooccipital area. Almost all of the dural sinuses, especially cavernous sinus and occipital sinuses were occluded with thrombus. Histopathological examination confirmed the widespread thrombosis. Toxicological tests didn’t reveal any toxic or abused agent.

There weren’t any thrombus in the pulmonary or crural veins in the both cases.

DISCUSSION

Thrombosis of the intracranial veins and sinuses presents with a remarkably wide spectrum of symptoms and signs, thus mimicking numerous other disorders [8]. Clinical features can be grouped into four different syndromes: (I) isolated intracranial hypertension, (II) focal neurological deficits, (III) encephalopathy and (IV) cavernous sinus syndrome [2, 6, 11].

Each of these four syndromes has distinctive clinical features. Headache with or without papilloedema and visual disturbances are observed in intracranial hypertension. Focal neurological deficits marked with hemi or monoparesis, while altered level of consciousness is found in encephalopathy. On the other hand, cavernous sinus syndrome manifests itself with oculomotor nerve palsies [6, 11].

Most frequent symptoms and signs reported by patients include headaches, seizures, and focal neurological signs, decreased level of consciousness and disturbances of vision (papilloedema).

These symptoms can present in isolation or in association with other symptoms which may or may not be related to the predisposing factors [4, 5, 6, 8, 12, 13]. Most frequently, DST presents with headache and this symptom was recorded in both of our cases.

Isolated thrombosis of the different sinuses and veins results in diverse clinical pictures. Ocular signs are dominant in cavernous sinus thrombosis while isolated thrombosis of the lateral sinuses present mostly as isolated intracranial hypertension. Left transverse sinus occlusion is usually followed by aphasia and clinical picture is frequently more severe with coma, mental troubles. Thrombosis of deep cerebral venous system presents with bilateral motor deficits [8].

In both of the presented cases, all of the sinuses were occluded by widespread thrombosis. Cavernous and sphenoparietal sinuses might be the origin in the first case. In this case, fractures on the basis of cranium due to closed head injury, was followed by meningitis and DST. Dural sinus thrombosis has a reported incidence of 4% after a penetrating head trauma [14].

Although common when skull fractures depressed or cross the sinuses, thrombosis can occur in association with mild head injuries even in the absence of fractures [8, 14]. Proposed theories, for occlusion of sinuses in head trauma include compression of the sinuses by intracranial edema or bleeding, intramural hemorrhages, extension of trauma from the scalp or injured emissary...
veins, and trauma to the sinus endothelial lining [1, 8, 14].

Hence, actual occurrence of cerebral venous sinus thrombosis in traumatic injuries might be higher than realized, and if overlooked fatal complications may develop [15, 16] as happened in our case.

Typical CT findings of cerebral venous sinus thrombosis include hyper attenuating thrombus in occluded sinus, the delta sign and cord sign on non-enhanced CT, and the empty delta sign on enhanced CT, which are present in up to 30% of cases [11].

Therefore, computed tomography can be inconclusive in some patients. It is a common mistake to depend entirely on the radiologist for a definite diagnosis and on the laboratory tests for an etiology [14, 15]. Similarly, the fracture and broken bone fragment and DST were not identified on the initial non contrast CT in our first case. Yuen et al. reported that diagnosis must rely on a high index of suspicion in children who sustained blunt head injury, and have persistent symptoms of giddiness and/or vomiting out of proportion as well as to the severity of the injury when there is not any obvious parenchymatous lesion on imaging studies [14]. Thus in our first case, cerebral, cerebrovascular events and cerebral venous sinus thrombosis should have been suspected in the first examination and the follow up.

In the second case, the patient’s nonspecific presentation such as diarrhea, fever and pharyngitis resulted in a missed diagnosis and death. In accordance, Shah reported that the initial signs and symptoms of cerebral venous sinus thrombosis can mimic more common disorders and may cause a delayed and missed diagnosis [11].

Therefore diagnosis of cerebral venous sinus thrombosis should be considered in all young and middle-aged patients with recent onset of unusual headache, stroke-like symptoms, and seizures, even in the absence of the usual risk factors for arterial thrombosis [15].

Sasidharan et al. recommend that, in order to avoid delay in initiating the specific treatment, patient should be managed as cerebral venous sinus thrombosis if the clinical picture is highly suggestive and consistent with or at least not against the diagnosis of cerebral venous sinus thrombosis, even if there is no other definite etiological diagnosis on CT or MRI [15].

Despite normal PT/INR and platelet count on the second case, total WBC and neutrophile, lymphocyte basophile, and monocyte counts were critically increased and CRP was very high. Bone marrow examination hadn’t been done and her medical records were received after several weeks of medico legal autopsy and blast level were unknown.

Thus, we couldn’t identify whether she had a leukemia or leukemoid reaction and whether she had hematologic or infectious disease. Because AST level was higher than ALT, it was suspected that there was tissue damage (other than liver) and blood hypercellularity was above critical level. Local or systemic infection, genetic or acquired thrombophilia, malignancies, and hematological disorders such as polycythemia are accepted as risk factors for DST [2].

DST should be suspected in patients with known predisposing risk factors, recent onset of unusual headache and neurologic symptoms especially if there is a head trauma [9, 16] as observed in the first case. Ferro et al. reported that female patients with more severe clinical presentations and patients in high-income countries tend to be diagnosed earlier. In the second case, patient had low income with a seasonal job in a farm and she also had a non-specific symptoms. Hence her diagnosis delayed considerably. As a consequence, a high index of suspicion is absolutely essential to diagnose cerebral venous thrombosis [15, 16].

In conclusion, diagnosis of DST is basically clinical and one should be aware of the varied clinical presentations of this condition and should get a CT or MRI as the next step for supportive evidence of cerebral vein and dural sinus thrombosis and to rule out other mimickers [15]. Otherwise, the case results in clinical missed diagnosis and physicians can be accused of malpractice as given by this report.

References