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CASE REPORT

Ruptured concomitant dermoid cysts of conus medullaris and cervico medullary junction

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ABSTRACT

Cranio spinal dermoid tumors are rare, benign, slow growing congenital, cystic uni or multi locular tumors, which arise from the inclusion of epithelial tissue within the neural grove during embryonic development.[1] In this case report, we present an uncommon presentation, of a case of concomitant dermoid cysts at conus medullaris and cervico medullary junction that ruptured into the central canal.

Key words: Fat saturated T1-weighted sequence, rupture, spinal dermoid

Introduction

Intramedullary spinal dermoid cysts, without any spinal dysraphism is rare. Only a few cases of dermoid cysts in medulla oblongata is reported. Presentation of dermoid cysts at multiple sites, in the cord, with rupture is not reported in literature. We report such a case in this article.

Case Report

A 21-year-old female patient presented with weakness of bilateral lower limbs, incontinence of urine and frequent urinary tract infection past 10 years. On clinical examination, bilateral lower limb paraparesis (2/5 on both lower limbs) and signs of renal failure like pedal edema, facial puffiness was present. On biochemical evaluation, her blood urea was 80 mg/dL and creatinine was 2.4 mg/dL suggestive of chronic renal failure. On ultrasound examination, her bladder was hugely distended with trabeculations, which was looking a neurogenic bladder. She was referred to our radiology department for craniospinal magnetic resonance imaging (MRI), to rule out any spinal etiology of her neurogenic bladder. The MRI examination was performed with a 1.5 T unit (Siemens Magnetom sympony MRI Machine). T1-weighted (T1W) (repetition time [TR]/echo time [TE], 400/9.2 ms) turbo spino echo (TSE), T2-weighted (T2W) TSE (TR/TE, 4200/96 ms) mutiplanar images were obtained. In addition, after intravenous (IV) Gd GTPA injection, T1W fat-suppressed axial and sagittal images were obtained.

A well-defined inverted tear drop shaped, T1W and T2W hyperintense expanding mass lesion was noted in the medulla oblongata, with extension to the cervical spinal cord up to C2 level. The hyperintense signal was suppressed, on fat-suppressed T1W sequences. In spinal MRI, another well-defined slightly lobulated and vertically elongated heterogeneous mass lesion observed on T1W and T2W images at the conus medullaris from L1 to L3 (lumbar) level with few hyperintense regions within it on T1W image, which was suppressed, on fat-suppressed sequences. Both the cervico medullary junction and the conus medullaris lesions showed no significant enhancement on post IV Gd diethylenetriamine penta-acetic acid MRI. Fat intensity, which filled the syrinx cavity inside the spinal cord was detected in the cervicothoracic level. Computed tomography (CT) screening of cervical spine and lumbar spine was done, which showed that both cervicomedullary and lumbar lesion contained, fat as internal contents (cervicomedullary lesion had mean Hounsfield units [H.U] −114.6 and lumbar lesion had mean H.U−31.6).

All these MRI findings, such as location of the mass, hyperintense signal intensity in T1-W MRI, nonenhancement, suppression on fat-suppressed imaging and observation of fat particles in the syrinx cavity were in accordance with a ruptured craniospinal dermoid tumors [Figures 1-3].

Cranial screening of the patient was done with MRI fluid attenuation inversion recovery axial and diffusion weighted sequences. No significant abnormalities detected in the cerebral hemispheres and cerebellum.

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The patient had near total resection of the cervico medullary tumor, by sub occipital craniotomy approach. On peroperative, the lesion looking like a brownish well-defined cyst, is located within the medulla and upper cervical cord. The cyst has a small rent in its lower pole. The cyst was subjected to histo-pathological examination. The specimen sent for pathological examination, showed a cyst which was lined by a stratified squamous epithelium, beneath which sebaceous glands, lobules of adipocytes, hair follicles, and vascularized fibrous tissue were identified, thus, confirming the preoperative radiological diagnosis of dermoid cyst [Figure 4].

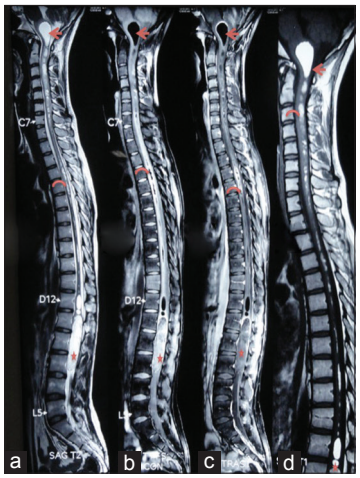

Figure 1: (a-d) Sagittal magnetic resonance imaging. Well-defined T1-weighted (T1W) (d) and T2-weighted (T2W) (a) hyperintense expanding mass lesion (arrow) noted at the medulla with extension to the cervical cord (up to C2) showing signal suppression on fat-suppressed T1W sequence (b). A T1W (d) and T2W (a) heterogeneous lobulated mass lesion (star) noted at the conus medullaris. Both these lesions show no significant enhancement on T1W contrast sequences (c) fat signal intensities (curved arrow) were noted within the syrinx cavities at cervicothoracic spinal cord regions (a-d)

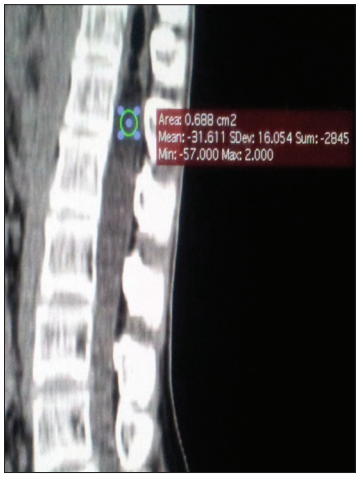

Figure 3: Noncontrast sagittal computed tomography imaging lumbar spine shows a expansible slightly lobulated mass lesion at the conus medullaris with fat contents (mean Hounsfield unit measures – 31.6)

The patient was under postoperative intensive care for about 2 weeks. The patient came after 40 days after hospital discharge, and her lower limb weakness has significantly improved from 2/5 to 4/5. However, her urinary symptoms showed no significant improvement. As she presented with urinary tract and respiratory tract infections currently, she was advised to come after 1-month to plan surgery for her conus lesion, after reduction of her infective symptoms.

Discussion

Dermoid cysts are congenital midline cystic tumors, representing about 20% of all intradural tumors seen during the 1st year of life. They generally present in patients younger than 20 years. Males and females are affected equally. The lumbosacral region is the most common site to be affected (60%).[2] Next most common site is cauda equina (20%).

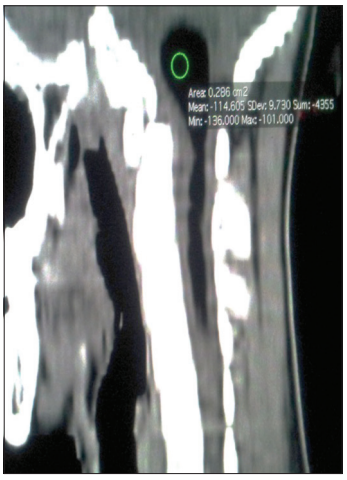

Figure 2: Noncontrast sagittal computed tomography imaging of cervical spine showing a inverted tear shaped expansible lesion at the medulla oblongata with extension to cervical spinal cord, with fat as internal contents (mean Hounsfield unit meaures – 114.6)

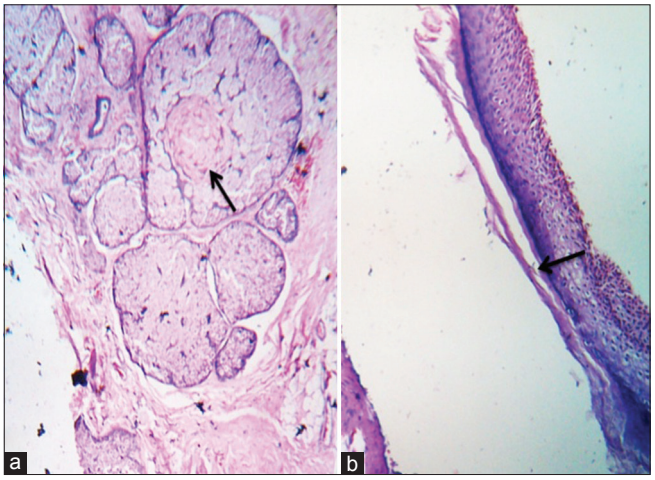

Figure 4: Histological analysis of the specimens (a) cellular keratin debris (arrow) and fat are seen (b) the cyst was covered by keratinized squamous epithelium (arrow) (H and E, ×10)

They are rarely found in the cervical or thoracic spine. 20% of those lesions are associated with a dermal sinus tract.[3] May be associated with occult spinal dysraphism.

They result from the inclusion of epithelial tissue within the neural groove during embryonic development. They have a lining of stratified squamous epithelial cells, containing sebaceous glands, sweat glands and hair follicles, which account for the fat intensity signal seen in these lesions.

Spinal dermoid cysts are often asymptomatic, and if symptomatic, motor disturbances, pain, sensory disturbances, and bowel or bladder dysfunction may be present[4] as in this case. If ruptured, they may become acutely symptomatic, which is associated with a high morbidity and mortality.[5] A dermoid cyst can rupture during surgery, after a trauma or spontaneously.

They have variable imaging appearances, but commonly appear as a mass of cerebrospinal fluid (CSF) density/intensity with fat density/intensity components. On CT examination, it presents as a well-defined mass, isodense to CSF, often with hypodense components (fat), with minimal enhancement. Widening of the spinal canal, flattening of the pedicles and laminae and osseous erosions may be demonstrated. Calcification may be seen.

On MRI, Signal intensity may be homogeneous or heterogeneous. In T1W imaging, the cyst may be hypo or hyperintense (hypointensity due to water content/ hyperintensity due to the presence of fatty secretions of sebaceous glands). In T2W imaging, the cyst is usually hyperintense, with no enhancement or mild rim enhancement with no obvious diffusion restriction. If the rupture occurs, multifocal T1W high signal areas (fat) are demonstrated within the subarachnoid space and/or ventricular system.

Surgical excision is the treatment of choice, which can improve or stabilize the symptoms in the majority of patients.[6]

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