INTESTINE INTO SACRAL CANAL A RARE FINDING IN MARFAN SYNDROME. CASE REPORT

El Mostarchid Brahim∗,1, Mamoune El Mostarchid∗, Mohcine Salami∗, Asri Abad Chrif∗ and Miloud Gazzaz∗

∗ Department of Neurosurgery, Mohammed V Military Teaching Hospital of Rabat Morocco.

ABSTRACT Background: Dural ectasia is a well-described feature of the Marfan, which is considered a major criterion in the diagnosis of Marfan Syndrome. Anterior sacral meningocele can be asymptomatic or symptomatic. The symptoms are mostly related to surrounding organs affected by mass effect. Case Description: Here, we present a case of anterior sacral meningocele with digestive structures inside the sacral canal via bone defect with unusual radiological findings. Conclusion: intestine structure into sacral canal with anterior meningocele must be kept in mind in the syndrome Marfan.

KEYWORDS Marfan syndrome, Anterior sacral meningocele, MRI, intestine in sacral canal

Case report

A 40-year-old woman presented for bilateral sciatica and chronic back pain. She was known to have a confirmed Marfan syndrome according to the revised Ghent nosology with ophthalmological and cardiac complications. She reported episodic abdominal pain and constipation. Neurological examination showed abolition of Achilles reflexes. Somatic examination showed a Marfanoid habitus with arachnodactyly and ligamentous laxity. Spinal lumbar CT scan showed enlargement of sacral foramina associated with heterogeneous content (Fig 1. A). Lumbosacral MRI demonstrated a huge intra sacral mass. This masse was heterogenous and was identified as an enteric structure with his meso (Fig1. B.C. D). The dural sac is pouched and compressed. She received symptomatic medical therapy with good results. With two years of follow up, she is paucity symptomatic. Surgical excision was not performed considering a stationary picture and neurological status after two years since diagnosis. Recently a family has reported that she presented sudden death 4 years later, probably by a cardiac complication. Unfortunately, no medical documents were available.

Discussion

Marfan’s Syndrome (MFS) is a connective tissue disorder, mainly involving the cardiovascular, musculoskeletal, and ocular systems. MFS diagnosis is made according to the revised Ghent nosology [1,2,3]. The sacral abnormalities in Marfan syndrome are attributed to defective collagen resulting in expansion of weakened dura, which leads to pressure erosion of contiguous osseous structures.

Our patient presented a large anterior sacral defect with the digestive structures pushed into. This finding is very rare. We were unable to find similar published radiological findings. We think that the high abdominal pressure has pushed the visceral continent into the sacrum for the second time. These findings are very rare and must be considered in MFS. Dural ectasia is defined as dilation of the dural sac and/or the nerve root sheaths, which was considered a major criterion in diagnosing MFS [1,2,3], due to its low frequency in the general population and association with some specific disorders, such as MFS. For example, it occurs in 95% of adult patients with Marfan syndrome and over 40% of children with this syndrome.

It’s known that the severity of dural ectasia increases with age. Anterior sacral meningocele results from dural ectasia due to subarachnoid space enlargement by cerebrospinal fluid (CSF) pulsation. Higher continuous pulsatile pressure of the CSF in the lumbosacral area with the patient standing would progressively affect a weakened and congenitally defective dural wall and spinal canal [3,4,5]. Clinical presentation of anterior meningoceles is highly variable. Small or late-onset isolated meningocele is usually asymptomatic. Early phase symptoms are mostly re-
which are exacerbated when standing up.

Neurological symptoms, such as low back pain, headache, pain and paresthesia in lower limbs, genital and rectal pain, abdominal pain have been described. Anterior sacral meningoceles have been reported to enlarge with age, and complications can be related to mass effect, including constipation, urinary retention, and prolonged labor in women.

A sacral meningocele can also predispose the patient to recurrent episodes of meningitis. Although very rare, fistulous communication of an anterior meningocele with adjacent bowel has been reported, usually resulting in bacterial meningitis. Kangan-Rivet et al. [6] reported an unusual case of E. coli meningitis in a patient with MFS.

Patient’s with marfan’s syndrome are at risk for meningitis from gut pathogens, especially E coli from meningeal abnormalities acting as a portal of entry. In addition, surgical options should be explored for patients with recurrent meningitis from colonic pathogenst with meningoceles or other anomalies.

Syndrome of intracranial hypotension is rare but known related to complications of MFS. CSF leak is the only proven cause, either iatrogenic such as a lumbar puncture or spontaneous, for the occurrence of intracranial hypotension syndrome and secondary neurological symptoms [3]. On the other hand, dural ectasia and intracranial hypotension syndrome are little known complications of Marfan Syndrome. Rahimizadeh et al. [9] reported an exceptional anterior sacral meningocele complicated by rectothecal fistula and rectorrhea. Same JD et al. [4] reported the first case of diverticulitis complicated by fistulous communication of sigmoid colon with anterior sacral meningocele in an anterior sacral meningocele in the MFS.

Spinal lumbosacral CT scan on bone view showed enlargement with the remodelling of the posterior wall of the vertebral bodies of the lumbosacral canal. A full-spin MRI in sagittal, axial and coronal views demonstrated the anterior meningocele as dilatation thecal sac. The compression or fistulous communications between digestive or urinary structures cab be demonstrated. Digestive radiological explorations are necessary if a symptom of digestive compression are presented. Differential diagnoses of dural ectasia include congenital arachnoid cysts and intradural tumours that widen the spinal canal. When dural ectasia extends to the pelvis as an anterior sacral meningocele, it can look like a pelvic mass.

The management can be only conservative in asymptomatic patients. Surgery can be discussed in symptomatic or complicated cases. Management of anterior spinal meningoceles was discussed by Wang et al. [7]. He advises that management often requires precise treatment based on the different conditions of each patient. Surgical intervention has been proposed for the treatment of symptomatic anterior spinal meningoceles. The goal of surgery is to safely disconnect the linkage between the cyst and CSF from subarachnoid space to prevent further enlargement of the cyst or reaccumulate cystic fluid. In the cases of digestive fistula, disconnection is necessary to prevent recurrent meningitis. Recurrences unknown aetiology must be considered possible digestive fistula with anterior meningocele in patients with MFS.

**Funding**

This work did not receive any grant from funding agencies in the public, commercial, or not-for-profit sectors.

**Conflict of interest**

There are no conflicts of interest to declare by any of the authors of this study.

**References**

1. Loeys BL, Dietz HC, Braverman AC, et al. The revised Ghent nosology for the Marfan syndrome. J Med Genet 2010; 47(7):476-85.
2. Sónia Gomes Coelho , Ana G Almeida. Marfan syndrome revisited: From genetics to the clinic. Rev Port Cardiol. 2020; 39(4): 215-226.
3. Pichott Andrea, Bernstein Tomás, Guzmán Guillermo, Faría Guillermo, Aguirre David, Espinoza Aníbal. Dural ectasia and intracranial hypotension in Marfan syndrome. Rev. chil. Pediatr. 2020; 91(4): 591-596.
4. Same JD, Johnson PT, Horton KM, Fishman EK. Diverticulitis complicated by fistulous communication of sigmoid colon with anterior sacral meningocele in a patient with Marfan syndrome. Radiology Case Reports. (Online) 2012; 7:442.
5. Böker T, Vanem TT, Pripp AH, Rand-Hendriksen S, Paus B, Smith HJ, Lundby R. Dural ectasia in Marfan syndrome and other hereditary connective tissue disorders: a 10-year follow-up study. Spine J. 2019; 19(8):1412-1421.
6. Kangath RV, Midturi J. An unusual case of E coli meningitis in a patient with Marfan’s syndrome. BMJ Case Rep. 2013, 5; 2013

7. Yue Long Wang, Bin He, Jinhao Yang, Jianguo Xu, Jiagang Liu, Siqing Huang. Individualized management of giant anterior meningoceles case series. Medicine (Baltimore). (2020) 99:14.

8. Maconi G, Pini A, Pasqualone E, Ardizzone S, Bassotti G. Abdominal Symptoms and Colonic Diverticula in Marfan’s Syndrome: A Clinical and Ultrasonographic Case Control Study. J Clin Med. 2020, 28; 9(10):3141.

9. Rahimizadeh A, Ehteshami S, Rahimizadeh A, Karimi M. Anterior sacral meningocele complicated by rectotheal fistula and rectorhea: A case report and review of the literature. Surg Neurol Int 2020; 11 :117.