MATURE TERATOMA OF THE CONUS MEDULLARIS : EXCEPTIONAL PRESENTATION IN AN ADULT PATIENT

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ABSTRACT : Teratomas are germinal cell tumors, affecting mainly patients of the pediatric population. The Spinal topography and the age of presentation; are extremely rare parameters in this case. This report is about an adult male, who presented intestinal transit disorders and lower limb weakness for two weeks. The radiological investigation revealed an intramedullary spinal tumor at the level of the conus medullaris. After surgical resection, the histological studies were concomitant with a mature teratoma. We analyzed all the characteristics of the case and we presented a brief review of the literature about similar cases.

Key words *: Teratoma, conus medullaris, Adult.*

INTRODUCTION

Teratomas represent a group of germ cell tumor, that contains the three embryonic layers; mesoderm, ectoderm and endoderm. The pineal and suprasellar regions are the most affected regions in the central nervous system. The histological differentiation is the key factor to grade this tumor into mature, immature or malignant teratoma [1, 2].

The spinal topography for these tumors is extremely rare with a reported prevalence of 0.15-0.18 % of all spinal tumors3. For the pediatric population, 5-10% of spinal neoplasms are teratomas [2, 4, 5]. This rate is much lower in adults compared to infants; as it was reported in several works [2,6,7].

In this report, we present a case of an intramedullary mature teratoma at the level of the conus, affecting an adult patient. The clinical history is also detailed and differences are discussed. Similar cases reported in the literature are reviewed and resumed in the present work.

CASE REPORT

History and clinical status:

A 31 years old male, was referred to our department with a several weeks history of lower limb weakness and diffuse parethesias. This course of symptoms worsened 10 days before the admission, by intestinal transit disorders for which he was treated also before the radiological investigation. The physical examination revealed a paraplegic patient presenting saddle anesthesia and complaining of diffuse paresthesias in his legs .The patient presented also urinary retention, for which a catheter was placed and finally a localized hypertrichosis at the level of L1.

Radiological evaluation:

The lumbar CT showed an isodense lesion within the vertebral canal, extending from D12 to L2. Some indirect features were present in the bony windowing of CT. a significant widening of the vertebral canal diameter from D12 to L1 (26.1mm). We noticed also a scalloping of the posterior wall of the vertebral bodies at the same level. (Figure 1).

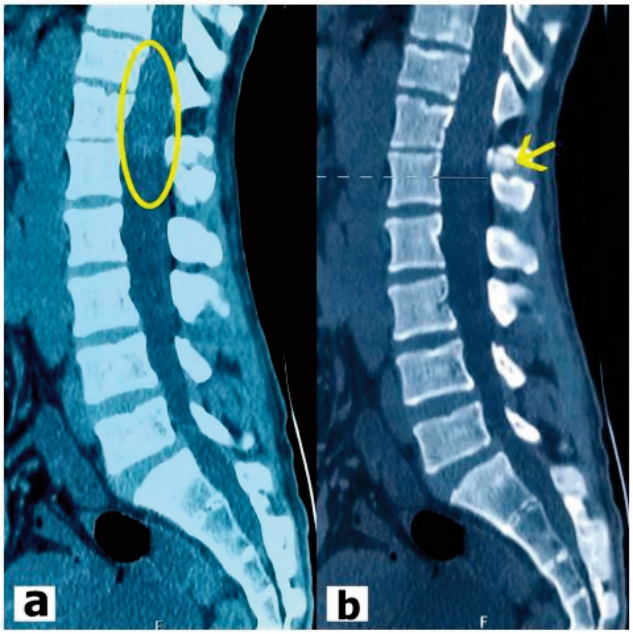

Figure 1 : lumbar CT in sagittal plans .(a): presence of tumoral mass at extending from D11 to L1 ,within the vertebral canal .it appears slightly hyperdense .(b): bony windowing that shows a widening of the canal, and presence of a bony fragment a the level of L1.notice the scalloping of the posterior walls of vertebrae at the level of the lesion .

The spinal MRI showed an intramedullary neoplasm, oval shaped within the conus medullaris . The measures were; 18×20 mm for diameter and 34 mm for the length extension. The tumor appears hyperintense in T1 and T2 sequences. In T2 FAT SAT sequences, the signal stays hyperintense .This feature allows us to differentiate it from a lipoma or a melanoma. (Figure 2)

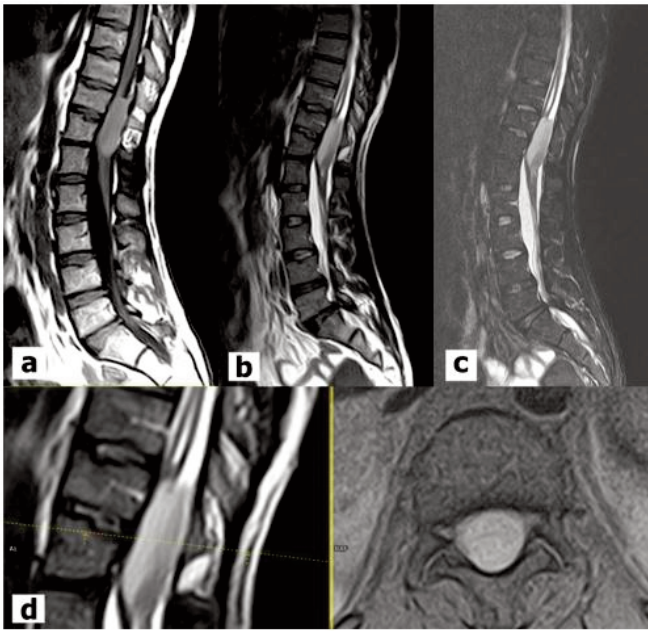

Figure 2 : Spinal MRI of the patient on sagittal and axial plans ,T1,T2,T2Fat Sat. (a): T1 sagittal section demonstrating a hyperintense intramedullary lesion at the level of the conus medullaris measuring 18×20mm .(b): T2 sagittal section :the lesion is moderately hyperintense and heterogeneous with an exophytic extension reducing then the subarahnoidian space at this level . (c): T2 FAT SAT sequence :the tumor is mainly hyperintense .the contents of the tumor were not of a lipomatous nature .(d): sagittal and axial sequences showing the intramedullary topography of the tumor within the conus medullaris .

SURGICAL MANAGEMENT AND HISTOLOGICAL STUDIES:

Given the patient's clinical condition, an emergency surgery was performed. Before the procedure, a full lumbar X-ray with a radiopaque landmarks (D12 – L1) to plan the right position of skin incision.

After completing a subperiostal dissection of the paravertebral muscles, the spinous process and laminae were exposed from D10 to L2 than resected .

A midline durotomy was performed. At this level, we noticed an abnormal yellowish color of the subarachnoidian space. Further microsurgical dissection, revealed an exophytic medullary cyst that presents a transdural bony attachement at the level of L1.

The cyst was made of pink wall and a green liquid. After aspirating a thick material inside the cyst , we performed a gentle dissection of the wall from the conus medularis . The aim of dissection at this level was to achieve maximal tumoral resection and preserve the conus .In some instances, the surgical resection was not possible to perform because of the significant adhesions between the cyst and the medullar tissue.

After a full microscopic analysis of the surgical field, the hemostasis was performed , using hemostatic agents, than the dura was closed in a watertight fashion. The same work was done for the rest of the other plans. The immediate postoperative course was uneventful.

After few days, we noticed a significant changing of the motor status. He recovered partially and the patient was able to move using a walker with the physiotherapist. Another important fact was the improvement of the saddle anesthesia. A partial sensory functions improvement was reported by the patient in his legs.

The residual symptom for the patient was diffuse paresthesias provoking a disturbing insomnia. The medical management was made of Pregabalin ; 200mg /day and Amitriptyline. The evolution after this treatment was favorable.

The histological studies were concomitant with a mature teratoma. The detailed report revealed the presence of a multi-layered keratinized squamous epithelium and a cystic formation is lined with a ciliated pseudo-startified cylindrical epithelium, the underlying chorion is the seat of bundles of smooth muscle fibers. To appreciate more this data, we have illustrated the microscopic histological appearance of mature spinal teratoma treated by Oktay & al 8. The histopathological examination of these lesions demonstrates a tissue composed of fully differentiated cells without the presence of malignant components. (figure 3).

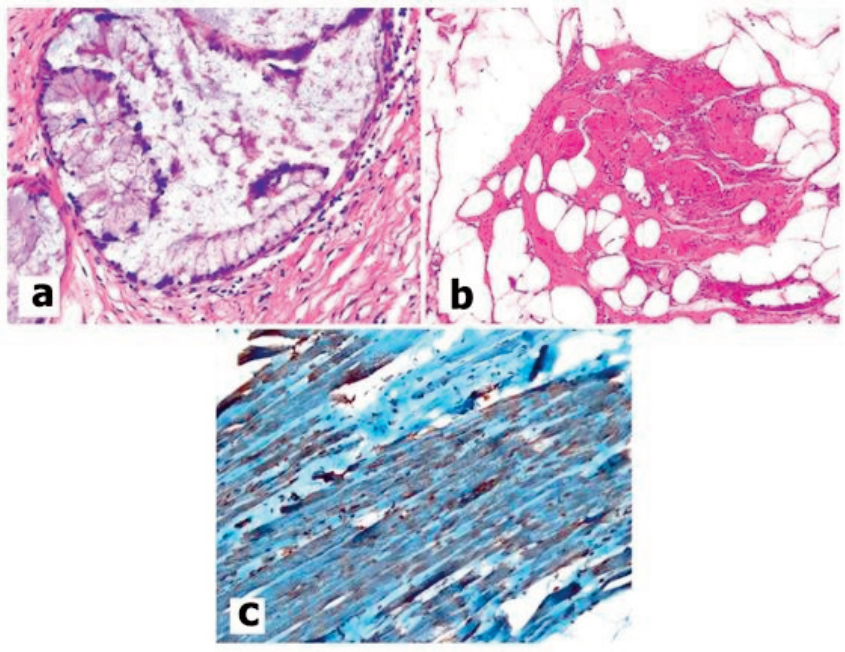

Figure 3: Histological appearence of a similar mature teratoma from Oktay & al work's8.
(a) Photomicrograph (H and E, ×40) showing connective tissue containing mucinous epithelium (b) photomicrograph (H and E, ×20) showing adipose tissue containing mature muscle cells (c) photomicrograph (IHC, ×40) showing smooth muscle cells reacted positively with actin.
Our policy was to evaluate clinically the patient one month later and radioligically with a lumbar MRI every 6 months during the first year.

The first imaging (figure 4) after surgery was performed after 8 months and revealed a small remnant of the cyst's wall at the level of the conus .(ring enhancement in the axial section (d)).

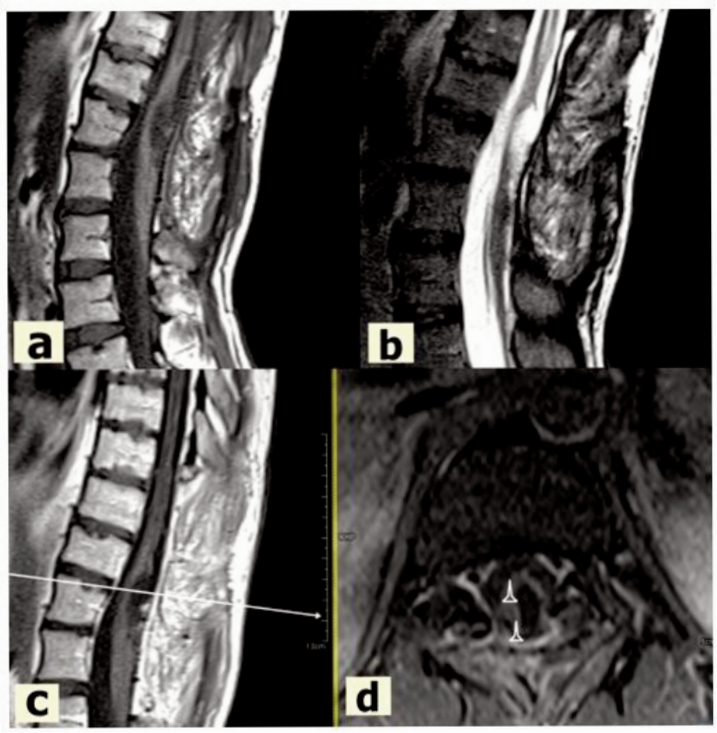

Figure 4 : MRI imaging (8months after surgery) in several sequences ;(a) :sagittal T1 sequence,(b) : sagittal T2 ,(c):sagittal T1 +contrast with level of section (white line).(d): axial FAT SAT + contrast sequence. All images show gross total resection of the teratoma without intensity changement of the conus . We can notice in (b) image, the persistence of syringomyelia above the resected lesion. Image (d): irregular enhancement inside the conus (stars) is the persistent wall of the cystic part of the tumor.

DISCUSSION

Teratomas are rare lesions affecting mostly the pediatric population, and could occur in the whole of the central nervous system(CNS). The spinal localization and especially the conus medullaris level are both very exceptional features [9]. Spinal teratomas account around 0.2-0.5 % of all spinal tumors [9-11].

The congenital aspect of their pathogenesis makes their onset in adults more uncommon, reducing then the number of reported cases in the literature. Two main theories were raised to elucidate the genesis of these tumors; the desembryogenic theory and the misplaced germ cell theory [12-15]. In the first theory (desembryogenic), the pluripotent cells are the origin of spinal teratoma. The occurence of chaotic differenciation for this pluripotent cells in a disturbed environment leads to the constitution of spinal teratoma [12,15]. In the second theory (misplaced germ cells), these pluripotent cells presents an abnormal postion during the migration from the yolk sac to the gonad. This failed process leads to the genesis of teratoma [14].

According to previous reports [16-18], the special feature in the adult patients presenting spinal teratoma is the scarcity of bony anomalies of vertebral bodies ; often encountered in children. Although our patient presented a lumbar cutaneous hypertrichosis ,the lumbar CT didn't show any bony malformation as spina bifida . The sagittal section of lumar CT showed an abnormal bony density between laminae . This finding was confirmed during surgery, where a bony tract in the epidural space was found at the level of the cystic part of the lesion.

Most of spinal teratomas are revealed by the appearance of sensory disorders such diffuse numbness or in other circumstances; a progressif weakness of the lower limb. These symptoms could be accompanied by pain in some patients [16, 17, 19]. In our case, the special additional symptom to the classic presentation was the occurrence of intestinal transit disorders. The compression

of the conus medullaris is the most plausible explanation for the transit disorders.

MRI evaluation remains the most valuable technique for these tumors. There is not specific feature for these lesions and they exhibit a mixed signal depending on the contents (bone, fat) [11]. The CT is important in detecting solid parts, associated anomalies, and plan the bony trajectory of the surgical approach. Several findings were found in CT in patients harboring spinal teratoma, we can mention; spina bifida [20], vertebral bodies fusion 11 and diastomatomyelia [21].

Our patient presented a cystic teratoma at the level of the conus with a hyperintense signal in all sequences and kept also the same signal in T2 FAT SAT sequences proving the non lipomatous nature of it .the lumbar CT demonstrated indirect features of the lesion; a widening of the vertebral canal, scalloping of the posterior wall of vertebral bodies and a bony fragment at the level of L1.

The histological grading of these lesions is another important parameter that should be considered during the management. Teratomas are classed into mature, immature and malignant group. For the mature group, they contain fully differentiated germ cells. In the composition of immature tumors group, a fetal tissue is found during microscopic studies. The third group of teratoma is the most aggressive because of the presence of a malignant component in histological studies. The histological analysis must cover the whole of the operative specimen to define the real group of teratoma . This procedure is performed to detect a possible malignant component of the tumor and not miss the true grading of it [22].

Management of teratoma is maximal microsurgical resection with preservation of neurological functions. In our case the cyst's wall, was not totally removed from the posterior aspect of the conus medullaris, because of the existence of intimate adhesions between the two tissues . This finding is also encountered in some other works [23-25], and the same precautions were considered about the extension of resection.

For totally or partially malignant teratomas, adjuvant radiation therapy should be completed for patients. The histology in our case was concomitant with mature teratoma which is the benign form. Periodic imaging and clinical evaluation were considered for the patient in the post operative management. We reviewed the literature about reports of conus medullaris teratoma, with an analysis of the following parameters: associated anomalies, resection, outcome and the residual symptoms in the postoperative period (Tab 01).

Study /year	Number of cases	Associated anomalies	Resection	Outcome	Residual symptoms
Nicoletti et al, 1994[26]	1	Caudal exophy of the conus	Incomplete	Stationary	Same to pre operative
Caruso et al, 1996[27]	1	Absent	complete	Improved	/
Al-Sarraj et al, 1998[13]	1	Absent	Incomplete	/	/
Fan et al, 2001[28]	1	Tuft of hair, congenital ventriculus terminalis, syrinx	Complete	Improved	/
Caruso and Colonnese, 2006[29]	1	Absent	Partial	Improved	Urinary disturbances
Kahilogullari et al. 2006[30]	1	Absent	Complete	improved	/
Mohindra et al, 2008[31]	1	Absent	Complete	Improved	/
Ijiri et al, 2009[32]	1	Absent	Complete	Improved	/
Benes et al, 2009[33]	1	Tuft of hair	Partial	Improved	/
Jian et al, 2010[34]	2	Absent	Complete	Improved	/
Turan et al, 2016[35]	1	Absent	Complete	Improved	Residual pain
Ernest Lekgabe et al, 2017[36]	1	Absent	Partial	Improved	/
Eleni Triantafyllidi et al, 2018[37]	1	Absent	Complete	Improved	Partial hypoesthesia
Present case	1	Lumbar tuft of hair	Partial	Improved	Residual pain

Table 1: reported cases of the literature of conus medullaris teratomas . (/: not reported).

The table shows that associated anomalies with this rare tumor are not frequent, and are represented mainly by a cutaneous tuft hair, which is the case of our patient. The microsurgical resection of conus teratoma remains partial in several works .this limitation is due to the existence of intimate adhesions between the lesion and the conus medullaris, making then the occurrence of postoperative neurological deficit more important in total resections. This concern was also encountered during our surgical procedure.

In almost cases, we notice an improvement of the preoperative clinical status of patients as it is in the present report. Residual pain was only reported by Turan et al but in fact it was a sensory change in the heel and gluteal region of the patient's foot. The intensity of pain was significantly more important in our patient, but had a favorable outcome after medical treatment.

CONCLUSION

Conus medullaris teratoma is a rare presentation in a daily neurosurgeon activity, and remains even exceptional in the adult population. Surgery is the modality of choice in the management of these lesions and the histological grading is an important feature for predicting the prognosis In most cases. Total resection in most of cases is not feasible because of the intimate adhesions of the tumor to the functional neural tissue of the conus. Although the previous limit, a significant percentage of patient harboring this tumor improve clinically in the postoperative course.

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