CASE REPORT

Primary dural-based MALT lymphoma mimicking falx meningioma: a case report

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Abstract

Primary dural lymphoma (PDL) is very rare and often misdiagnosed as meningioma based on neuroimaging characteristics.

A 36-year-old woman presented with headache, behaviour changes and ataxia. She had no history of oncology or immunodeficiency. Magnetic resonance imaging showed a solitary mass lesion in the anterior and middle thirds of the falx cerebri with bilateral growth and a moderate mass effect, which was interpreted as a falx meningioma. Gross total resection of the lesion involving the dura mater of the falx cerebri was performed by bilateral parasagittal craniotomy. Surprisingly, histological examination showed no meningothelial components. Instead, there was diffuse lymphocytic and plasma cell infiltration. Immunohistochemistry was positive for B-cell antigens. The final histopathological diagnosis was primary dural mucosa-associated lymphoid tissue lymphoma. Systemic screening examination was then performed and did not reveal additional tumor foci or lymphadenopathy. Follow-up brain MRI at 3 months did not reveal tumor progression. Neither chemotherapy nor radiotherapy were given, and the patient is under observation.

Diagnostic errors can cause delays in pathogenetic treatment of primary dural lymphoma. Thus, PDL should be considered in the differential diagnosis of meningiomas

Keywords: MALT, marginal zone lymphoma, central nervous system, dura mater, MRI, case report

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Introduction

Primary dural lymphoma (PDL) represents less than 1% of all lymphomas of the central nervous system and less than 0.1% of all non-Hodgkin lymphomas (1). The mean age of diagnosis of PDL is 50-55 years old, and it is believed that it is more common in female patients (2). The prognosis of PDL is relatively benign and can be treated by surgical resection, with or without subsequent radiation therapy (3). However, because of its rarity, it is often misdiagnosed as meningioma based on radiological characteristics (4), while diagnostic errors can cause delays in pathogenetic treatment. Thus, PDL should be considered in the differential diagnosis of meningiomas.

Neuroimaging alone cannot provide the final diagnosis (despite the presence of some relatively specific signs, there is always a diagnostic challenge), and detailed clinical information is very important. Unfortunately, it is believed that PDLs usually occur in patients without a notable clinical history (5).

Different treatment regimens exist in patients with PDLs without consensus in this area (3), and the main treatment is surgery followed by chemo- or radiotherapy. However, it has been shown that in the case of solitary dural lesions without bone invasion and/or CSF dissemination, total surgical excision of the tumor could be the only treatment option (1).

Because it is a very rare disease, information from case reports can be useful for neurosurgeons, as well as radiologists and oncologists. We present a case of primary dural MALT lymphoma that mimicked falx meningioma in a young woman.

Case report

A 36-year-old woman presented with headache, behavior changes and ataxia. She had no history of any kind of immunodeficiency or chronic neurological, hematological, infectious, or autoimmune diseases.

Magnetic resonance imaging (MRI) showed a contrast-enhancing solitary mass lesion in the anterior and middle parts of the falx cerebri with bilateral growth and the "dural tail" sign (***Figure 1***).

There was minimal edema of the peritumoral brain tissue and no signs of invasion; there was slight intrinsic signal hyperintensity in the precontrast T1-WI (Figure 1b) and very low apparent diffusion coefficient (ADC) values (approximately 400*10-6 mm2/sec, Figure 1d); however, there were no signs of hypoperfusion according to ASL data (CBF value was approximately 45 ml/100 ml/min, Figure 1e).

These neuroimaging findings were interpreted as consistent with meningioma. Thus, the preoperative diagnosis was a falx meningioma, and no additional examinations were performed.

The surgery was performed under general anaesthesia by bilateral parasagittal craniotomy using a bilateral interhemispheric approach ***(Figure 2***).

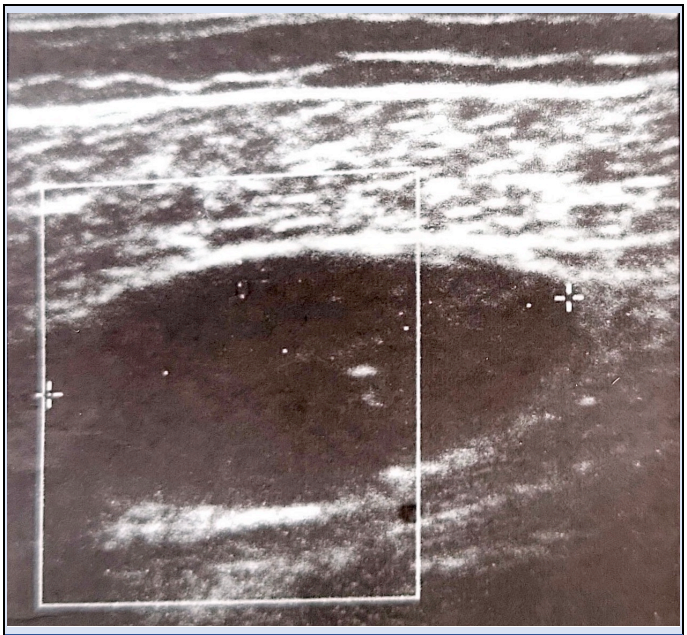

Figure 1. Lower leg soft tissue ultrasound demonstrating well circumscribed oval lesion in the region of the middle third of the calf muscles

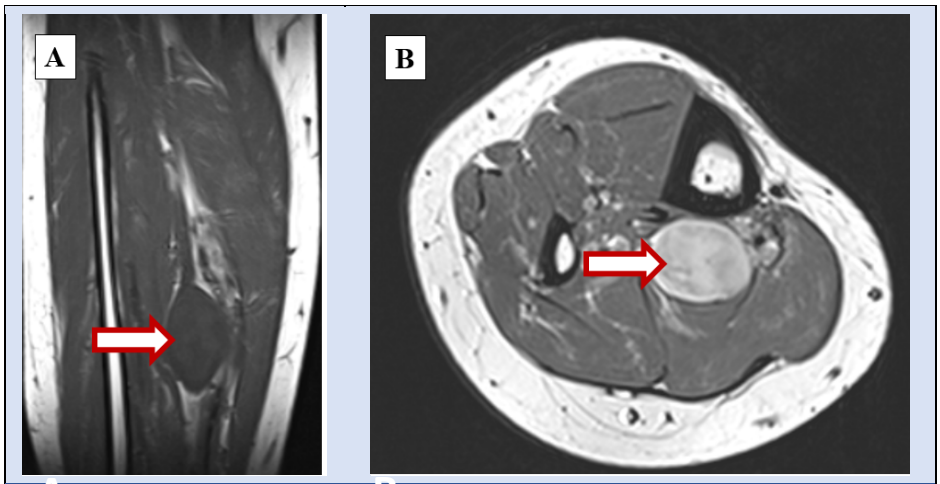

Figure 2. MRI of the right leg showing encapsulated oval shaped lesion within the intermuscular space in the posterior aspect of lower leg in the curse of tibial nerve. Tumor showed homogenous isointense on T1 (A) and heterogeneous hyperintense signal on T2 (B) MRI sequence. Arrows are pointing at the lesion.

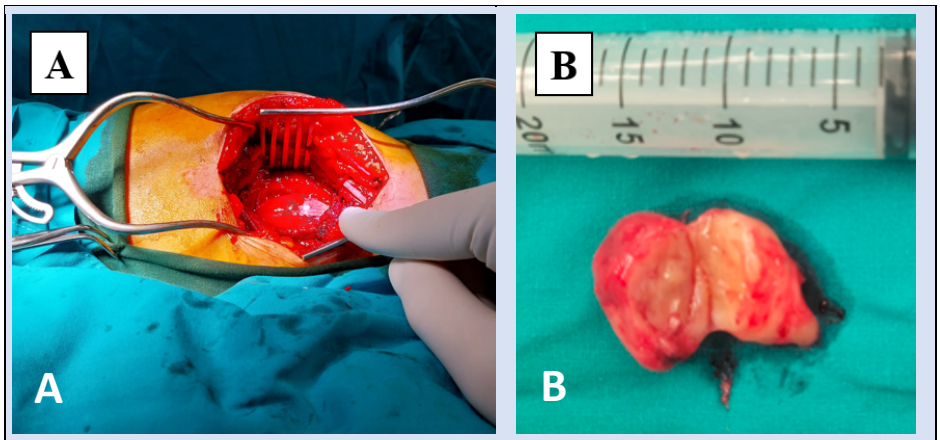

Figure 3. Intraoperative view of tumor. (A) after (B) cut in half after removal in one piece

Patient was operated under general anesthesia with microsurgical technique. After incising the epineurium of tibial nerve, tumorous lesion measuring about 3x4 cm in diameter was removed after meticulous preparation of surrounding nerve fascicules. This intracapsular approach was chosen, which enabled the removal of the tumor without additional damage to nerve function, e.g. the principle implies enucleation with gentle opening and longitudinal tumor capsule incision, separating the surrounding nerve fascicles under large magnification using operative microscope (***Figure 3***).

Fortunately, there was no disruption of tibial nerve continuity, and tumor was completely removed. Postoperative period was uneventful, and the patients was discharged home on the 3th postoperative day. Histopathological examination of the lesion confirmed presumed diagnosis of ancient schwannoma (***Figure 4***).

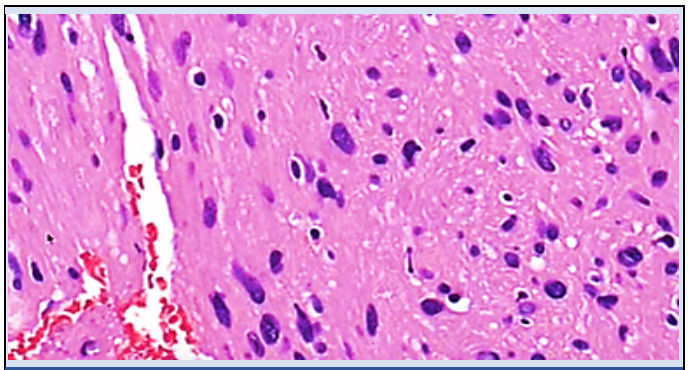

Figure 1. Histopathology indicative of the ancient schwannoma

The patient was regularly followed up at timely intervals for one year since the excision of the lesion. The patient was without recurrence of tumor during the follow-up period, and she was completely pain free immediately after operation, without neurological deficit.

Discussion

Schwannoma is a peripheral nerve sheet slow growing tumor, also known as neurilemoma, neurocytoma, peripheral gliomas, and neurinoma. It is the most common peripheral nerve tumor, and usually is presented as an asymptomatic, slow-growing, solitary, firm, well-circumscribed, encapsulated and palpable tumor (3). They are usually find in the head, neck and spine, while other locations are rare (4). Schwannomas are most frequent in adults between the ages of 30 and 60, and they affect both men and women equally. Most schwannomas are asymptomatic however they can cause functional deficits and pain depending on which nerve is affected. Most common peripheral nerve sheet tumors are schwannomas, followed by neurofibromas, hybrid nerve sheath tumors and perineuriomas (5). Although schwannoma represents a rare entity with incidence of about 5 cases per 100,000, they are the most common type of peripheral nerve tumors in adults. On the other hand, malignant peripheral nerve sheath tumor is a rare and aggressive type of sarcoma with incidence reported to be 1 in 100,000 adults per year, accounting for about 5 to 10% of all soft tissue sarcomas. We present a 35-years-old female patient with schwannoma in the rare location, between deep calf muscles involving the main trunk of the tibial nerve.

Schwannomas cause a varied symptomatology from asymptomatic phase to severe phase of nerve involvement with wide spectrum of clinical symptoms such as parasthesias, hypoesthesia, tingling, motor weakness, and positive Tinel's sign (6). Although, chronic leg pain can be caused by variety of causes, detailed anamnesis, and precise clinical and neurological examination can significantly narrow the differential diagnosis (2). Perineurioma symptoms differ from other peripheral nerve sheet tumors, presenting as a mononeuropathy of slow onset and gradual progression, resulting in neurological deficits like hypoesthesia or, much more frequently, a muscular weakness but, rarely with a positive Tinel's sign. In about 60% of the other patients, a Tinel's sign was triggered by tapping. This corresponds with the data in the literature that the Tinel's sign is highly indicative for a peripheral nerve sheet tumor and should lead to further diagnostics (7). In our patient peripheral neuropathy with positive Tinnel sign in the region of the medial portion of the right calf was present.

Among all radiological modalities, MRI is a gold standard for the diagnosis of peripheral nerve sheet tumors. MRI enhanced the preoperative diagnosis of nerve sheath tumors and provided information regarding the tumor characteristics, such as size, precise location, and relationship with surrounding structures. Nevertheless, the clinical preoperative diagnosis of schwannoma is often difficult because of its slow growing nature, high mobility, soft tissue texture, sometimes painless nature, and clinical symptoms that are shared with other conditions. Although a precise diagnosis is not possible only on the basis of radiological images, the MRI findings can indicate differences between malignant peripheral nerve sheath tumors and benign tumor lesions, especially neurofibromas based on several factors such as the largest dimension of the tumor mass, peripheral enhancement pattern after contrast administration, type of perilesional edema, and presence of cystic component in the tumor (8). Schwannomas may be misdiagnosed as fibromas, gangliomas, lipomas, or xanthomas (9). Phalen reported accurate diagnosis of schwannoma in one third of upper extremity schwannoma patients (10). Although peripheral nerve sheath tumors are relatively rare entity, general practitioners and doctors of all specialties must suspect the presence of such tumor as one of the possible diagnoses in the case of body or limb tumor. The time to diagnosis (TTD) as a factor in disease prognosis is increasingly in the focus of clinicians and is an important factor in the prognosis of many diseases (11). Also, complete failure of therapy should raise the suspicion of a possible other diagnosis. In the study of Uerschels et al. prolonged conservative treatment was present in 28.9% with peripheral nerve sheet tumors, resulting in pronounced neurological deficit in two patients and neuropathic pain syndrome in 5 patients (7). Our case is interesting since patient had symptoms for more than 10 years, and for many years she was treated for suspected lumbar disc herniation. This case illustrates that a properly taken medical history and a profound clinical examination cannot be replaced by any other diagnostic test, and apart from fallowing standard guidelines for clinical

practice, personalized medical approach is still *conditio sine qua non*.

Treatment for schwannoma is determined by the location of the tumor, if it is causing pain, and how rapidly it grows. Sometimes observation is indicated, but surgery is essential if symptoms occur or suddenly worsen, or when malignant alteration is suspected, which was also one of the most important reason for our patient's surgery, since she had a significant increase in pain in the last few months. Main goal is enucleating the tumor while avoiding nerve damage with internal neurolysis, so usage of operative microscope is strongly recommended (6). In most of the cases schwannomas are eccentrically located and do not involve the main nerve trunk, but tend to displace the nerve fibers peripherally in contrast to neurofibromas, which grow within the nerve fibers, potentially necessitating the partial or complete resection of nerve fibers, resulting in nerve deficit (7). In our case all nerve fibers were preserved by meticulous preparation, and this finding was also indicative of schwannoma. After performed operation our patient didn't have any postoperative complication nor neurological deficit.

Conclusion

Schwannoma is a peripheral nerve sheet tumor. Diagnosis is difficult when the tumor is in rare location, especially if the tumor is not palpable. However, a well-taken medical history and a detailed clinical and neurological examination are indispensable tools for making the right diagnosis. Also, in cases of radicular leg pain, one of the differential diagnoses should always be a peripheral nerve tumor...

Disclosures

Conflict of Interest: All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest (such as honoraria; educational grants; participation in speakers' bureaus; membership, employment, consultancies, stock ownership, or other equity interest; and expert testimony or patent-licensing arrangements), or non-financial interest (such as personal or professional relationships, affiliations, knowledge or beliefs) in the subject matter or materials discussed in this manuscript.

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References

1. Kumar R, Ranjan R, Jeyaraman M, Kumar S. Tibial Nerve Schwannoma: An Unexplained Cause of Lateral Foot Pain - A Rare Case Report and Review. J Orthop Case Rep. 2020 Dec;10(9):1-6.
2. Edwards PH Jr, Wright ML, Hartman JF. A practical approach for the differential diagnosis of chronic leg pain in the athlete. Am J Sports Med. 2005 Aug;33(8):1241-9.
3. Maraziotis T, Panagiotopoulos E, Panagiotopoulos V, Panagiotopoulos K. Neurilemoma of the popliteal fossa:Report of two cases with long subclinical course and misleading presentation. Acta Orthop Belg. 2005;71:496–9.
4. Han KJ, Lee YS, Park M. Digital nerve schwannoma of the hand. J Hand Surg Eur Vol. 2012 May;37(4):361-2.
5. Galbiatti JA, Milhomens GRDS, Bertozzo LG, Escames L, Milhomens Neto PA, Galbiatti MGP. Retrospective Analysis of 20 Patients Affected by Schwannomas in the Upper and Lower Limbs. Rev Bras Ortop (Sao Paulo). 2020;55(5):629-636.
6. Aleksić V, Prelić J, Radojević M, Ćulafić R. Schwannoma of the digital nerve. A rare case report. Romanian neurosurgery. 2021;35(4):490-492.
7. Uerschels AK, Dengler NF, Chihi M, Lenkeit A, Dinger TF, Jabbarli R, Sure U, Hagenacker T, Wrede KH, Gembruch O. Benign peripheral nerve sheath tumors: an interdisciplinary diagnostic and therapeutic challenge. Neurosurg Rev. 2023 Aug 18;46(1):205
8. Wasa J, Nishida Y, Tsukushi S, Shido Y, Sugiura H, Nakashima H, Ishiguro N. MRI features in the differentiation of malignant peripheral nerve sheath tumors and neurofibromas. AJR Am J Roentgenol. 2010 Jun;194(6):1568-74
9. Chelsea P, Ghazle H, Bhatt S, Dogra V. Schwannoma of the Tibial nerve. Journal of Diagnostic Medical Sonography. 2010;26(4):205-208.
10. Phalen GS. Neurilemomas of the forearm and hand. Clin Orthop Relat Res. 1976;114:219–222.
11. Launay E, Cohen JF, Bossuyt PM, Buekens P, Deeks J, Dye T, Feltbower R, Ferrari A, Kramer M, Leeflang M, Moher D, Moons KG, von Elm E, Ravaud P, Chalumeau M. Reporting studies on time to diagnosis: proposal of a guideline by an international panel (REST). BMC Med. 2016;14(1):146.