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Large multicystic spinal lesion in a young African migrant: a problem of differential diagnosis

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SUMMARY

We describe a rare case of large, fully cystic spinal schwannoma in a young adult from The Gambia. The initial clinical suspicion was spinal cystic echinococcosis. He came to our attention reporting progressive walking impairment and neurological symptoms in the lower limbs. An expansive lesion extending from L2 to S1 was shown by imaging (ie, CT scan and MRI). Differential diagnoses included aneurysmal bone cyst and spinal tuberculosis and abscess; the initial suggested diagnosis of spinal cystic echinococcosis was discarded based on contrast enhancement results. The final diagnosis of cystic schwannoma was obtained by histopathology of the excised mass. Cystic spinal lesions are rare and their differential diagnosis is challenging. Awareness of autochthonous and tropical infectious diseases is important, especially in countries experiencing consistent migration flow; however, it must be kept in mind that migrants may also present with 'non-tropical' pathologies.

BACKGROUND

Fully cystic lesions of the spine are rare. Differential diagnosis includes cystic degeneration of solid schwannomas, fully cystic schwannomas, aneurysmal bone cyst, cystic haemangioblastoma, cystic neurofibroma, cystic ependimoma, cystic meningioma, cavernous lymphangioma, bronchogenic or neurenteric cysts, dermoid and epidermoid cysts, and dural/arachnoidal cysts. Infectious causes of cystic lesion of the spine include bacterial abscess, tuberculosis and cystic echinococcosis.[1–4] We report the case of a man from The Gambia, who recently arrived in Italy through Libya and was found to have a fully cystic, large, slow-growing, multiloculated spinal lesion on CT scan and MRI of the spine. The initial suggested diagnosis was cystic echinococcosis of the spine, although neuroradiological images showed features which could unequivocally exclude this diagnosis. Histopathology was, however, needed for a definite diagnosis among non-echinococcal differential diagnoses.

CASE REPORT

In January 2019, a 23-year-old Gambian patient presented to the outpatient clinic for migrants at the Department of Infectious Tropical Diseases and Microbiology of the IRCCS Sacro Cuore Don Calabria Hospital, Negrar, Verona, Italy.

He had arrived in Italy in March 2017, after spending 6 weeks in Libya. Soon after his arrival, he was admitted in a primary care hospital in Southern Italy due to pulmonary tuberculosis, anaemia and scabies; during the admission, he was diagnosed with an ischiatic ulcer with exposed bone and meningocele. He was treated for scabies and anaemia; treatment for drug-sensitive pulmonary tuberculosis was also started. The patient reported that the ischiatic ulcer appeared in the absence of any trauma or injury, approximately 2 years before, and progressively worsened. In May 2017, the patient moved to the Veneto region in Northern Italy. In December 2017, he presented to the plastic surgery department of our hospital, due to worsening of the ischiatic lesion. The patient underwent surgical curettage and vacuum-assisted closure therapy; the histological examination of the removed tissue documented dermal sclerosis with associated inflammatory granulation tissue and angiogenesis; the presence of dysplastic or neoplastic elements was excluded. Then, he was regularly followed up with surgical debridement and wound dressings. Tuberculosis treatment was successfully completed in January 2018, with no signs or symptoms of relapse over time.

When he presented to our clinic in January 2019, he reported progressive walking impairment, weakness and pain in the lower limbs bilaterally. The physical examination revealed painful mobilisation of the lower limbs, associated with hypoesthesia and paraesthesia on the left side. He did not report fever, weight loss or night sweats.

INVESTIGATIONS

Laboratory tests showed a mild leucopenia (white cell counts 3300 cells/µL, with 28.3% neutrophils), which was already known and stable over time. Erythrocyte sedimentation rate and C reactive protein were within the normal ranges, as well as liver, kidney and thyroid function tests. He tested negative for HIV and hepatitis viruses. Chest X-ray and abdominal ultrasonography were unremarkable. A CT scan of the spine revealed an expansive, hypodense lesion, extending from L1 to S1 and to the psoas muscles bilaterally, causing marked vertebral erosion and scalloping. MRI of the lumbar and sacral spine was performed on a 1.5T MR imaging unit (Avanto, Siemens, Erlangen, Germany) using spine phased-array coils. After intravenous administration of gadolinium contrast material (gadoteridol 0.1 mmol/kg, 8 mL), 3D gradient recalled echo images (volumetric interpolated breath-hold examination, (VIBE)) were acquired in sagittal plane. The lesion was multiloculated, intradural with expansive behaviour and intraforaminal extension,

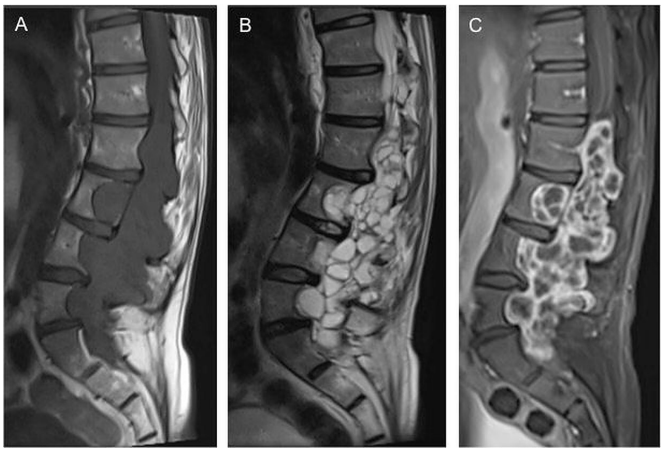

Figure 1 (A) T1-WI, sagittal plane, showing a lobulated, space-occupying mass with homogeneous hypointense signal intensity, extending from the L1 to the S1 vertebral level. (B) T2-WI, sagittal plane, showing an intrathecal high signal intensity mass with multiloculated appearance. (C) Gadolinium contrast-enhanced 3D gradient recalled echo (volumetric interpolated breath-hold examination) image, sagittal plane, showing a multiloculated mass with rim enhancement of the thick and irregular walls and intralesional septa after intravenous gadolinium administration. WI, weighted image.

scalloping the posterior wall of the lumbar vertebrae. The lesion was hyperintense on T2-weighted images (WI), homogeneously hypointense on unenhanced T1-WI, and did not show macroscopic fat content or areas with features suspicious for haemorrhage in different stages of evolution (figure 1A,B). The walls of the cysts and intralesion septa markedly enhanced on administration of gadolinium contrast medium (figure 1C).

Serology for cystic echinococcosis (Echinococcus IgG ELISA, DRG Instruments GmbH, Germany; and Cellognost Echinococcosis IHA, Siemens, Germany) was negative. The final diagnosis was achieved by histopathological examination of the surgically removed mass, which showed a well-demarcated tumour with large pluriconcamerate cystic and solid areas. The latter were composed of spindle cells arranged in fascicles, with immunohistochemical (IHC) reactivity for S100 and SOX-10, indicating likely origin from Schwann cells, and proliferative index of 2% as assessed by IHC for Ki-67. Based on the morphological and IHC findings, the lesion was diagnosed as a cystic schwannoma (cystic neurinoma), grade I, according to the WHO classification.[5]

DIFFERENTIAL DIAGNOSIS

Imaging and clinical features were compatible with a slow-growing, benign mass. Spinal cystic echinococcosis enters in the differential diagnosis of cystic spinal lesions. Indeed, although The Gambia is not endemic for cystic echinococcosis, the patient could have been infected during his travel through North Africa.[1] Also, Italy is endemic for cystic echinococcosis.[1] Although serology for cystic echinococcosis was negative, extrahepatic cystic echinococcosis, including echinococcosis of the bone, is often seronegative, making a negative serology inconclusive to rule out this infection. The diagnosis of spinal cystic echinococcosis, however, could be discarded on the basis of the presence of lesion wall enhancement shown by MRI on administration of gadolinium contrast medium. Other main differential diagnoses, characterised by compatible imaging, included aneurysmal bone cyst (although no evident, clear-cut typical fluid–fluid levels could be visualised) and infections such as spinal tuberculosis or abscess (although no clinical signs of bacterial infection were present).[3 4] Histopathology supported the final unexpected diagnosis of fully cystic spinal schwannoma.

TREATMENT

The patient underwent excision of the schwannoma, vertebroplasty and dorsosacroiliac (D11–D12–L1–S1) arthrodesis using transpedicular and iliac screws. After the neurosurgical procedure, he was transferred to the rehabilitation ward, where he carried out motor and neurological rehabilitation for a 6-month period. During this period, he wore an orthopaedic corset.

OUTCOME AND FOLLOW-UP

After surgery, the patient showed clinical improvement, achieving a good level of autonomy in daily activities (eg, dressing, eating and moving between bed and wheelchair). At the last follow-up in February 2021, his deambulation had further improved; notwithstanding, the need for a walking aid remained. A follow-up MRI will be scheduled according to the patient's availability.

DISCUSSION

Fully cystic lesions of the spine are rare. Schwannomas are benign neoplasms of the nerve root sheaths, most commonly occurring in the fourth–fifth decade of life in both sexes. They are the most common primary intraspinal tumours, generally with intradural extramedullary localisation; however, they are usually solid or heterogeneously solid masses localised at the cervical and lumbar level.[4 6] While cystic degeneration of solid schwannomas is well-described, predominantly or fully cystic schwannomas are uncommon, with only a few cases reported in the literature. Schwannomas generally show low-to-intermediate signal intensity on T1-WI and may be heterogeneous on T2-WI, with hyperintense areas corresponding to cystic portions.[4 7 8] Although there are no pathognomonic features of schwannoma on imaging, rim enhancement of the cystic portion maybe suggestive.[4] Similar characteristics may be evocative also of aneurysmal bone cyst and abscesses.

Aneurysmal bone cyst is a benign, highly vascular, multiloculated, locally aggressive osteolytic lesion involving most commonly the lumbar and sacral spine in children and adolescents.[3 9] In an aneurysmal bone cyst, the cyst's content shows variable signal, with a surrounding rim of low T1-WI and T2-WI signal, and the septa enhance on contrast administration.[9] Fluid–fluid levels are characteristic, but not exclusively observed in aneurysmal bone cyst, and may be absent in a proportion of cases.[3]

Spinal abscesses may be visualised as smooth, peripheral ring-enhancing lesions, hypointense in T1-WI and hyperintense in T2-WI, with central diffusion restriction in diffusion-weighted imaging (DWI). Although DWI was not performed in our case, clinical and laboratory findings did not support the hypothesis of a spinal abscess.

Cystic echinococcosis is caused by infection by the larval stage (metacestode) of the tapeworm parasite *Echinococcus granulosus* sensu lato. While the parasitic larvae in humans develop most commonly in the liver (≈70% of cases) and lungs (≈20%) as concentrically growing fluid-filled cysts, in the bones (involved in 0.5%–4% of cases) and the vertebral column (occurring in ≈50% of all cases of bony

involvement), the parasite growth is characterised by a slow, aggressive 'microvesicular' bone infiltration along trabecular spaces.[10] The lumbar spine is commonly affected in spinal cystic echinococcosis, often with ≥2 vertebral levels involved, and an extraspinal localisation is not always concomitantly present.[10] Imaging is the basis of the diagnosis of cystic echinococcosis.[11] However, while echinococcal cysts, especially in abdominal localisations, may show pathognomonic features on ultrasound, imaging features of cystic echinococcosis of the bone are non-specific, requiring differential diagnosis with neoplastic and inflammatory processes.[11] In our case, although The Gambia is not endemic for cystic echinococcosis, the patient could have acquired the infection in North Africa.[1] Serology for cystic echinococcosis was negative in two tests. However, even in the case of epidemiological and clinical suspicion, serology in bone cystic echinococcosis may be negative and therefore not diriment.[2] In our case, while the appearance of the lesion on unenhanced imaging could have been compatible with cystic echinococcosis, the marked enhancement of the lesion walls and septa on administration of gadolinium contrast medium ruled out this differential diagnosis, as *E. granulosus* cysts, being of parasite origin, by definition, do not enhance.

Other differential diagnoses (cystic haemangioblastoma, cystic neurofibroma, cystic ependimoma, cystic meningioma, cavernous lymphangioma, bronchogenic or neurenteric cysts, dermoid and epidermoid cysts, and dural/arachnoidal cysts) could be considered less likely on the basis of unenhanced imaging and contrast enhancement characteristics.[4 6 7 12]

Learning points

- Large cystic schwannomas are rare, benign neoplasms, which can be diagnosed by histopathological examination.
- Features of cystic echinococcosis on imaging should be widely disseminated and known, especially in endemic countries and in countries receiving migrants from endemic areas.
- Extrahepatic cystic echinococcosis, including echinococcosis of the bone, is often seronegative, making a negative serology inconclusive to rule out this infection.
- Awareness of autochthonous and tropical infectious diseases is important in an era of globalisation and intense travelling, especially in countries experiencing consistent migration flow; however, it must be kept in mind that migrants may also present with 'non-tropical' pathologies.

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