Benign notochordal cell tumour: Case report

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Case report

A 56 year-old woman, without antecedents, was examined for chronic lumbago. The X-rays of the lumbar vertebrae were normal. A CT scan and then an MRI were carried out.

The lumbar spine CT scan (Fig. 1) revealed a median posterior corporeal lesion on L5 of 36 mm in diameter. It was compressed, irregular and had several lytic zones. In MRI, the lesion (Fig. 2) was in hypersignal T2, hyposignal T1, non-enhanced. It complied with the cortex of the posterior vertebral wall, the posterior vertebral arc, the epidural space and the soft peri-vertebral tissue.

The most likely diagnosis was that of a benign tumour deriving cells from the notochord (BNCT). Considering their rarity and the difficult differential diagnosis with a chordoma, in particular, a vertebral biopsy under fluoroscopic guide was carried out with a Larédo-Hamzé coaxial tocard.

The histological examination (Fig. 3) of the biopsy samples revealed, within the spongy tissue, in the intertrabecular tissue, a replacement of the haematopoietic tissue by pseudo-lipomateous tissue made of vacuolised cells with weakly eosinophilic cytoplasm. Cytonuclear atypia or myxoid stroma was not noted. The trabeculae in contact were slightly sclerotic; the limits of the lesion were distinct.

The immunohistochemical techniques (Fig. 4) revealed diffuse positivity of the pseudolipoblastic cells with anti-AE1/AE3 antibodies, protein S100 and vimentin.

The anatomopathological examination confirmed the diagnosis of BNCT. Monitoring through imaging was proposed.

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Discussion

Initially described under the name “notochordal hamartoma of intra-osseous origin” or “giant notochordal residue”, BNCT are rare intra-osseous lesions. They are found along the entire spine, most often cervical or lumbosacral, and are asymptomatic or revealed through moderate pain.

They evolve little clinically, are limited to the vertebral body, without extension to the soft tissue and are small (<4 cm). In the CT scan (exceptionally on X-rays), the bone structure is respected or slightly densified without osteolysis. In MRI, the lesion presents a T1 hypointense without enhancement and is hyperintense in T2 [1].

Histologically, they consist of cell areas of pseudo-fatty appearance, creating residual fatty islands or haematopoietic marrow at the contact. Pseudocystic spaces may be observed with weakly eosinophilic pseudocolloidian alcian blue and positive PAS. The tumour cells may include several positive and diastase-resistant PAS granules in their cytoplasm. The bone in contact is often densified with moderate osteosclerosis. The immunophenotype of the tumoral cells is identical that of the cells of chordomas. There is a co-expression of protein S100, vimentin and epithelial markers. In the absence of clinical information and imaging, they may not be diagnosed due to their pseudo-lipomatosus appearance.

The differential diagnoses in imaging are metastases and bone lymphomas. These tumours may condense the bone matrix but the association bone condensation—hyposignal T1—hypersignal T2—absence of enhancement after the injection of gadolinium is only found for BNCT [2]. Usually, the osteocondensing metastases are in hyposignal T1-T2, enhanced according to the degree of condensation; the lymphomas are osteolytic, variable in hyposignal T1-T2, enhanced (+ infiltration of the soft tissue).

The main differential histological diagnosis is chordoma [3]. The distribution of the chordomas is anatomic and the intra-osseous locations are identical. They are locally aggressive, responsible for osteolysis with extension to the
Figure 3. Histological examination (HE staining) that shows the replacement of haematopoietic tissue by pseudo-lipomatosus tissue consisting of vacuolised cells with weakly eosinophilic cytoplasm (with different zooms).

Figure 4. Immunohistochemical examination with anti-AE1/AE3 antibodies, protein S100 and vimentin that reveals a diffuse positivity of the lipoblastic cells.
soft tissue. The imaging detects the osteolysis and the extra-osseous extension. Their signal in MRI is identical that of BNCT but are enhanced [2]. Their treatment is surgical, with the risk of local recurrence and distant metastases.

The second differential histological diagnosis consists of notochordal vestiges. It involves notochordal residues exceptionally observed in the inter-vertebral discs of the spine or at the base of the skull. These very rare asymptomatic lesions are morphologically close to chordomas but without anomaly, mitosis or necrosis.

BNCT and chordomas do not share the same prognosis since BNCT are most often painless lesions not requiring their removal but simple clinical and radiological monitoring [2]. The microscopic examination of needle biopsies can help distinguish them [1]. In 2010, Nishiguchi [4] first described the histological association of BNCT—chordoma. Is there a filiation between these two lesions or is there a chance association? This case report notes the importance of the collaboration between the radiologist and the pathologist since the MRI appeared to be that of a chordoma (invasion of the peri-vertebral soft tissue and enhancement). Monitoring by imaging should be systematic [5].

The diagnosis of BNCT should be familiar to pathologists, radiologists and surgeons in order to avoid heavy and useless surgery.

Conclusion

BNCT are recently described entities that, in reality, group an important nosology. Their filiation with chordomas still needs to be demonstrated. The distinction of these two entities is necessary since their treatment and prognosis differ. Needle biopsies provide a sure diagnosis. They are an alternative to surgical biopsies and radical treatment.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References