Indeterminate Dendritic Cell Tumor in Thoracic Spine: A Case Report

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BACKGROUND: **Indeterminate dendritic cell tumor (IDCT) is an extremely rare hematologic disorder with poorly understood pathogenesis. Occasionally encountered by hematologists, unusual presentations of IDCT have not been reported in the spine literature.**

METHODS: **We report a 51-year-old man who presented with a 3-month history of progressively worsening axial thoracic back pain radiating to his sides. Magnetic resonance imaging revealed a 3-cm enhancing mass at the T9 vertebral body with an exophytic component causing significant canal stenosis. Initial percutaneous biopsy revealed histiocytic sarcoma.**

RESULTS: **The patient underwent exploratory thoracotomy and en bloc resection of the lesion with T8-10 fusion. Final pathology results revealed IDCT with fibrosis. IDCT immunostaining was partially positive for Langerhans cell marker (positive for S100 and CD1a, but lacked Birbeck granules and Langerin stain) and partially positive for blastic plasmacytoid dendritic cell neoplasm. Additionally, it was positive for CD45, CD68, and CD163. Lymphadenopathy was absent in this patient.**

CONCLUSIONS: **Although first reported in the 1980s, IDCT has been omitted from most classifications owing to its rarity. Hematologists have debated the cell of origin; it is believed to comprise pre—Langerhans cells, as Birbeck granules are acquired after migration to the epidermis. IDCT remains of indeterminate origin. We report the first case of spinal IDCT. Familiarity with the histologic features is warranted to ensure accurate diagnosis and appropriate treatment.**

INTRODUCTION

Indeterminate dendritic cell tumor (IDCT), comprising so-called dermal indeterminate cells, is an extraordinarily rare hematologic neoplastic disorder.[1,2] First noted by Wood et al. in 1985, IDCT involves an altered homing mechanism of proliferation of antigen-presenting dendritic cells (DCs).[1] Because of its rarity, the etiology of IDCT is unknown, although the most frequently discussed possibilities are extrinsic stimulations and association with low-grade B-cell lymphoproliferative disorder.[1,3] The pathophysiology is not completely understood, and this often leads to misdiagnosis.

IDCT can develop in any individual with no particular predilection for age or sex.[2] The most common area of manifestation of IDCT is the skin because these cells predominantly reside in the skin and lymph nodes.[4] The skin of the trunk and extremities is usually affected.[5] There are 3 main types of DCs in the human body that have been described: dermal DCs expressing CD1a, dermal DCs expressing CD14, and Langerhans cells expressing CD1a.[5] Rowden et al.[6] suggested that indeterminate cells, originating from the same progenitor cell, are pre—Langerhans cells that fail to complete their acquisition of Birbeck granules while migrating toward epidermis. Therefore, the major difference between these DCs is the presence of ultrastructure Birbeck granules.[1] In addition, on immunohistology, IDCT cells stain CD1a and S-100 protein but not CD207 (langerin).[1] We describe our experience with a case of IDCT in the thoracic spine compared with skin and lymph nodes cases found in the past literature. Familiarity with the histologic features of IDCT is essential in spinal surgery to ensure accurate diagnosis and appropriate treatment of this rare entity.

MATERIALS AND METHODS

A 51-year-old man presented with a 3-month history of progressively worsening axial thoracic back pain radiating to his sides.

Key words
- Birbeck granules
- Epidermotropism
- Histiocytic sarcoma
- Indeterminate dendritic cell tumor
- Langerin

Abbreviations and Acronyms
DC: Dendritic cell
IDCT: Indeterminate dendritic cell tumor
LCH: Langerhans cell histiocytosis

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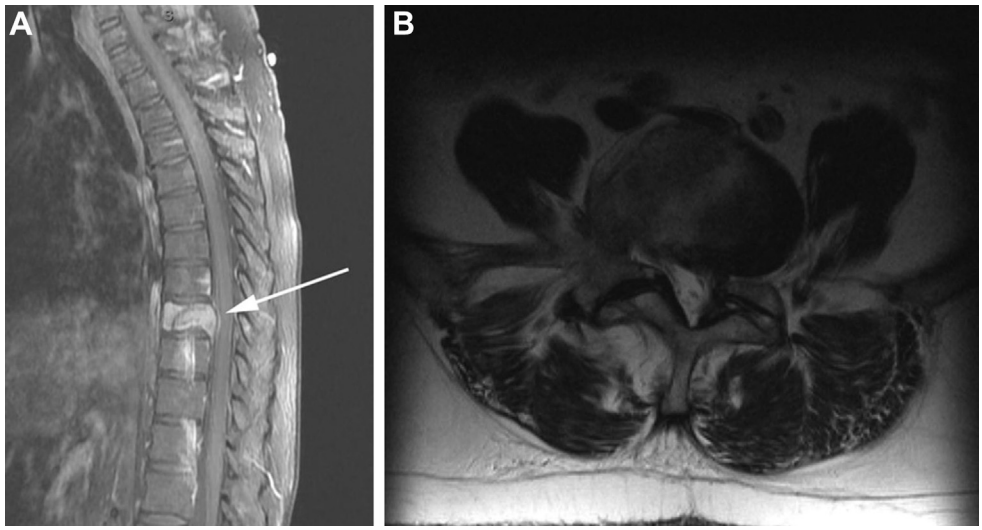

Figure 1. (**A**) Magnetic resonance imaging of thoracic spine reveals a 3-cm lesion (*arrow*) in the T9 vertebral body causing retropulsion and compression of the central canal. (**B**) Sagittal section at T9 vertebral body.

The pain was not associated with paresthesia or weakness. The patient denied any cutaneous or systemic symptoms. No lymphadenopathy was noted. Extensive serologic laboratory work-ups were normal.

Magnetic resonance imaging revealed a 3-cm enhancing mass at the T9 vertebral body with an exophytic component causing significant canal stenosis with epidural extension (Figure 1). Computed tomography angiography of the chest showed the presence of an artery of Adamkiewicz at the level of the lesion. Subsequent whole-body positron emission tomography scan confirmed low-level fluorodeoxyglucose-avid bilateral cervical subcentimeter nodes, likely reactive. Initial percutaneous needle biopsy showed histiocytic sarcoma.

The patient underwent a right exploratory thoracotomy and en bloc resection of the lesion with T8-10 posterolateral instrumented fusion using an expandable titanium cage and rib autograft. The seventh rib was also partially resected.

RESULTS

Histopathologic examination of the 4.5 × 3.7 × 2.1 cm resected mass revealed IDCT with fibrosis, chronic inflammation, and reactive woven bone formation. The mass was an irregularly shaped lytic tumor-like lesion with no clear laterality. It mainly involved the middle and posterior aspects of the vertebral body and extended through the cortex into the adjacent soft tissues. All inked resection margins were negative, and the clearance to the closest soft tissue margin was <0.1 cm.

The neoplasm was unusual and was composed of a multinodular proliferation of large histiocyte-like cells with hyperlobulated nuclei that had smooth to irregular contours (Figures 2 and 3). The cytoplasm of tumor cells was eosinophilic and abundant. The tumor cells demonstrated limited cytologic atypia, minimal mitotic activity, and no necrosis. The lesional cells were admixed with macrophages with significant lymphoblastic infiltration and surrounded by reactive fibrosis (Figure 4).

Immunohistochemistry showed that the lesion cells were positive for CD1a, CD45, CD68, and CD163 (pale) and focally expressed Oct-2 (dim nuclear staining) and S-100 (Figure 5). The cells stained negative for langerin, CD30, CD3, CD20, CD21, CD23, CD35, kappa and lambda light chains, tryptase, keratin, CK8/18, EMA, SALL4, BRAF, and desmin. The proliferation rate estimated from the Ki-67 stain was <2%.

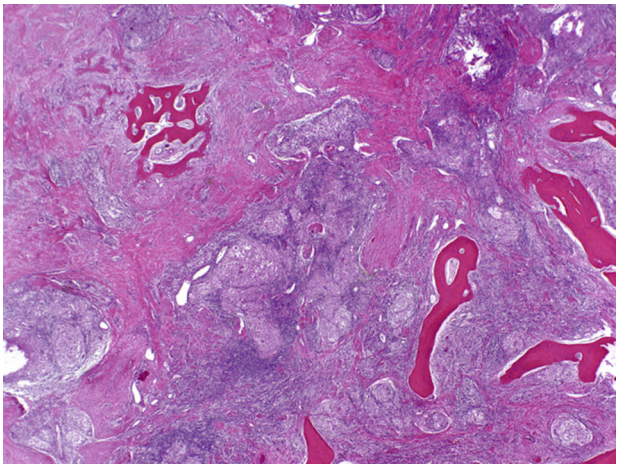

Figure 2. Cancellous bone destroyed by cellular infiltrate (magnification, 2×).