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TEST YOURSELF: ANSWER

Soft tissue mass at the infrascapular fossa

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Answer

Epstein–Barr virus (EBV)-associated smooth muscle tumor at the infrascapular fossa.

The EBV, a member of the herpesvirus family, is highly prevalent within the human population. It has been implicated in the pathogenesis of hematological, epithelial, and mesenchymal tumors. EBV-associated smooth muscle tumors (EBV-SMT) are related to an immunocompromised or immunosuppressed state.

Soft tissue tumors occurring secondary to an immunocompromised state were not recognized before 1970 [1]. It was only in 1995 that the association between smooth muscle tumors and EBV was established in patients who had undergone organ transplantation [2, 3]. These lesions are rarely diagnosed less than 3 months post-transplant and have been reported 29 years after renal transplantation [4]. They have an unusual predilection to occur in sites that have little smooth muscle, such as the brain or dura. Other sites of involvement include the adrenal gland, lung, gallbladder, bone, bladder, spleen, thyroid, and heart [5]. Multiple sites of involvement can occur in up to 68% [5]. In such cases, clonal analyses suggest that they arise from separate infection events rather than a metastatic phenomenon [2].

Radiologically, EBV-SMT frequently present as a non-specific enhancing soft tissue mass. Appearance can range from a vividly enhancing mass [6] to a more heterogeneous enhancing lesion [7]. MRI is sensitive to the detection of these tumors, but is not specific. The key to narrowing the diagnosis is an immunodeficient or an immunocompromised state, as in this patient.

A complete consensus on criteria for malignancy in deep smooth muscle tumors has not been established, but most experts suggest that the presence of cytologic atypia and mitotic activity (>1 mitotic figure per 50 high power fields) should raise suspicion of malignancy [8–10]. EBV-SMT, which often demonstrates mitotic activity and primitive round cell areas that may be mistaken for atypia, could potentially lead to a misdiagnosis of leiomyosarcoma. Multifocality in EBV-SMT may further suggest metastasis and thus malignancy. The prognosis of leiomyosarcomas of somatic soft tissues is generally poor, with approximately 50% 5-year survival, often influenced by tumor size. However, the prognosis of EBV-SMT (though based on relatively few cases) is usually good and related to the immune status of the patient rather than tumor size [5]. Consequently, recognition of this entity is crucial to the patient.

Epstein–Barr virus smooth muscle tumors are a rare entity. Other conditions that might be considered in the differential diagnosis of this case include a peripheral nerve sheath tumor (PNST), given the close proximity to the spinoglenoid notch where the suprascapular nerve is located. There are, however,

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no features of denervation within the infraspinatus muscle such as may accompany suprascapular nerve involvement [11]. Extramedullary relapse of ALL is also a concern because of the patient's medical history. The main sites of extramedullary relapses of acute leukemia are the central nervous system and skin, although leukemic infiltrates may relapse at other sites including the neck, spine, limbs, and body cavity [12].

However, in view of the clinical context of an immunodeficient or immunocompromised state, EBV-associated smooth muscle tumor should also be entertained.

References

1. Pritzker KP, Huang SN, Marshall KG. Malignant tumours following immunosuppressive therapy. Can Med Assoc J. 1970;103:1362–5.
2. Lee ES, Locker J, Nalesnik M, et al. The association of Epstein-Barr virus with smooth-muscle tumors occurring after organ transplantation. N Engl J Med. 1995;332:19–25.
3. McClain KL, Leach CT, Jenson HB, et al. Association of Epstein-Barr virus with leiomyosarcomas in children with AIDS. N Engl J Med. 1995;332:12–8.
4. Ferri L, Fraser R, Gaboury L, et al. Epstein-Barr virus-associated pulmonary leiomyosarcoma arising twenty-nine years after renal transplantation. J Thorac Cardiovasc Surg. 2003;126:877–9.
5. Deyrup AT, Lee VK, Hill CE, Cheuk W, Toh HC, Kesavan S, et al. Epstein-Barr virus-associated smooth muscle tumors are distinctive mesenchymal tumors reflecting multiple infection events: a clinicopathologic and molecular analysis of 29 tumors from 19 patients. Am J Surg Pathol. 2006;30:75–82.
6. Kumar S, Santi M, Vezina G, Rosser T, Chandra RS, Keating R. Epstein-Barr virus-associated smooth muscle tumor of the basal ganglia in an HIV+ child: case report and review of the literature. Pediatr Dev Pathol. 2004;7:198–203.
7. Gallien S, Zuber B, Polivka M, et al. Multifocal Epstein-Barr virus-associated smooth muscle tumor in adults with AIDS: case report and review of the literature. Oncology. 2008;74:167–76.
8. Weiss SW. Smooth muscle tumors of soft tissue. Adv Anat Pathol. 2002;9:351–9.
9. Hornick JL, Fletcher CD. Criteria for malignancy in nonvisceral smooth muscle tumors. Ann Diagn Pathol. 2003;7:60–6.
10. Miettinen M, Fetsch JF. Evaluation of biological potential of smooth muscle tumours. Histopathology. 2006;48:97–105.
11. Tirman PFJ, Feller JF, Janzen DL, Peterfy CG, Bergman AG. Association of glenoid labral cysts with labral tears and glenohumeral instability: radiological findings and clinical significance. Radiology. 1994;190:653–8.
12. Byrd JC, Edenfield WJ, Shields DJ, Dawson NA.. Extramedullary myeloid cell tumors in acute nonlymphocytic leukemia: a clinical review. J Clin Oncol. 1995;13:1800–16.