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TEST YOURSELF: ANSWER

A 22-year-old man with a posterior left shoulder mass

Joy Li[1] · Stephen P. Winter[1] · Rishabh Gattu[1] · Kyle D. Perry[2] · Kara Gaetke-Udager[1] · Eman Abdulfatah[2] · Jonathan B. McHugh[2] · Kristine E. Konopka[2] · Mohamed Abdelmohsen Bedewi[3] · Steven B. Soliman[1]

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Answer

Extraneural soft tissue perineurioma.

Discussion

Perineurioma (PN) is a rare type of benign peripheral nerve sheath tumor, first described in 1978 [1]. PNs are classified as intraneural; confined within a nerve, or extraneural; in the soft tissue or skin [2–4]. Extraneural PNs are further subdivided into soft tissue (most common), sclerosing, and reticular subtypes [2, 3]. Briefly, sclerosing PNs are classically found in young patients' hands [2, 5], and reticular PNs are characterized by extensively myxoid composition [3, 6].

PNs are composed of neoplastic cells with similar immunohistochemical features to normal perineural cells [2, 7]. Perineural cells normally make up the perineurium, which is a protective barrier between the epineurium and endoneurium of individual nerve fascicles [6, 7].

PNs have a wide age range, peaking in the middle age, and with a slight female predilection [3]. Although typically benign, malignant PNs have been rarely reported [3]. Unlike other peripheral nerve sheath tumors, PNs are unique in that there is no strong association with neurofibromatosis [7]. However, there have been some reports of PNs in both neurofibromatosis types 1 and 2 [8–10].

Similar to this case, extraneural soft tissue PNs present clinically with many benign features including being a well-circumscribed, firm, elastic, and slow-growing mass which can be mobile (Fig. 1). Furthermore, these usually do not affect range of motion, strength, or cause pain. On the contrary, intraneural PNs may lead to slowly progressive mononeuropathy or other neurologic deficits including weakness [2–4].

As seen in our case, radiographs may demonstrate a mass-like density without osseous involvement and rarely with mineralization/calcifications (Fig. 2). However, in a case series by Broski et al., they identified mineralization in 3 out of 7 extraneural PN cases that had CT or radiographic correlation [3]. On ultrasound, extraneural soft tissue PNs typically appear as a smoothly marginated, hypoechoic mass without significant vascularity on Doppler [5] (Fig. 3). On MRI, and similar to our case, PNs demonstrate T1-weighted hypointense to isointense signal, T2-weighted hyperintense signal, and demonstrate heterogeneous or solid enhancement [2, 3, 5] (Fig. 4). Extraneural PNs typically have no intralesional hemorrhage, surrounding edema, or associated muscle denervation [2, 3]. Extraneural intermuscular or intramuscular PNs can have a thin rim of peripheral fat [3], however, in our case of an infraspinatus intramuscular PN we did not appreciate this finding (Fig. 4). Extraneural PNs do not have an apparent nerve of origin, whereas intraneural PNs appear as fusiform enlargement of the affected nerve [3].

Based on imaging features, the differential includes other peripheral nerve sheath tumors, such as neurofibroma or schwannoma. However, neurofibroma and schwannoma

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✉ Steven B. Soliman
ssoliman@med.umich.edu

[1] Department of Radiology, Division of Musculoskeletal Radiology, University of Michigan, Ann Arbor, MI, USA

[2] Department of Pathology, University of Michigan, Ann Arbor, MI, USA

[3] Department of Internal Medicine, Prince Sattam Bin Abdulaziz University, College of Medicine, Al-Kharj, Kingdom of Saudi Arabia

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can have a target-like enhancement on MRI and may demonstrate intralesional hemorrhage or calcification [2, 3, 5]. Additional malignant considerations include soft tissue sarcomas and malignant peripheral nerve sheath tumors, although these often have more irregular margins and faster growth [2, 3, 5].

Since there are overlapping features, biopsy is needed for confirmation. In our case, an ultrasound-guided biopsy was performed and demonstrated a bland neoplastic cell population with variable cellularity (Fig. 5). Nuclei varied from ovoid to spindled/epithelioid-like and while the background stroma was predominantly collagenous, there were focal areas of myxoid features. Cells showed areas of positive "linear-like" staining for GLUT-1 and CD34 (Fig. 5). Cells were negative for S100, MUC4, SOX10, SMA, desmin, STAT6, and MDM2. Findings were diagnostic of PN [7]. Surgical excision is the treatment of choice and PNs do not typically recur after excision [2, 4]. The patient subsequently underwent marginal resection of the tumor and the diagnosis was confirmed.

PN is a rare and underrecognized tumor with characteristic immunohistochemical features. Knowledge of this entity and its imaging and immunopathologic features is important.

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Compliance with ethical standards

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Informed consent was obtained from the subject described in this report. Institutional review board protocol review was exempt per our institutional review board policies for this type of manuscript and since these examinations were clinically indicated. Our study complied with the Health Insurance Portability and Accountability Act.

Conflict of interest The authors declare that they have no conflict of interest.

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