Left Supraclavicular Swelling: Sclerosing Perineurioma

Antonio Tancredi[1,2], Paolo Graziano[3], Lucia Dimitri[3], Elvira Impagnatiello[4], Marco Taurchini[1]

Cite this article as: Tancredi A, Graziano P, Dimitri L, Impagnatiello E, Taurchini M.Left Supraclavicular Swelling: Sclerosing Perineurioma. Eurasian J Med 2018; 47-9.

[1]IRCCS "Casa Sollievo della Sofferenza" Hospital, Unit of General and Thoracic Surgery, Viale Cappuccini 1, 71013 San Giovanni Rotondo, Foggia, Italy
[2]IRCCS "Casa Sollievo della Sofferenza" Hospital, Department of Emergency, Viale Cappuccini 1, 71013 San Giovanni Rotondo, Foggia, Italy
[3]IRCCS "Casa Sollievo della Sofferenza" Hospital, Unit of Pathology, Viale Cappuccini 1, 71013 San Giovanni Rotondo, Foggia, Italy
[4]Azienda Sanitaria Locale di Foggia, "San Camillo De Lellis" Hospital, Unit of General Surgery, Via Isonzo, 71043 Manfredonia, Foggia, Italy.

Received: July 6, 2017
Accepted: December 1, 2017

Correspondence to: Antonio Tancredi

E-mail: antoniotancredi@virgilio.it

DOI 10.5152/eurasianjmed.2018.17214

ABSTRACT

A sclerosing perineurioma presents as a single asymptomatic papule or nodule located on the hands of adult patients; approximately 60 cases have been reported in medical literature since 1997. Histologically, it originates from the perineural cells of the peripheral nerves and presents epithelial membrane antigen (EMA) positivity and S100 protein negativity. Here, we present the case of a 58-year-old male admitted to our surgery unit complaining of left supraclavicular swelling of 1-cm in size, having no significant past medical history. A lymph node neck tumor was suspected, and the patient underwent surgery under local anesthesia in outpatient care. The intraoperative finding was a whitish mass, wooden-fibrous in consistency, strictly adhering to the skin and apparently fixed to the deep planes. Upon histological examination, a sclerosing perineurioma was diagnosed: neoplastic cells were immunoreactive for CD34, CD99, and EMA, and negative for S100 protein, smooth-muscle actin, pancytokeratin (AE1–AE3), CD31, neurofilaments, and beta-catenin. According to the benign biological tumor behavior, radical resection was considered an adequate treatment. Our case presents as peculiarity the unusual non-acral location (first reported as supraclavicular swelling) and the rare immunopositivity for CD34 and CD99.

Keywords: Sclerosing perineurioma, supraclavicular swelling, nerve sheath neoplasms

Introduction

Perineuriomas are uncommon, underrecognized, and mostly benign tumors originating from the perineural cells of the peripheral nerve sheath. First described by Lazarus and Trombetta in 1978, fewer than 150 cases have been reported to date in literature [1-4].

According to the anatomical site, they can be intraneural or extraneural. Intraneural perineuriomas are rarer; they typically affect young patients and involve the sciatic nerve and its branches and present with a progressive, painless motor deficit [5]. Extraneural perineuriomas are most common and can be divided into three subtypes: soft tissue, sclerosing, and reticular. They present as a single asymptomatic papule or nodule located on an extremity of the adult patient [2].

Here, we report our anecdotal experience of sclerosing perineurioma showing clinical and histological peculiarities which resulted in a challenging diagnosis for clinicians.

Case Report

A 58-year-old male with no significant past medical history was admitted to our unit complaining about a firm and painless swelling in the left supraclavicular region that slowly grown for 5–6 months to an overall size of 1 cm (Figure 1).

The preoperative blood tests and chest X-ray were normal, and because a lymph node neck tumor was clinically suspected, we did not consider it necessary to perform preoperative ultrasonographic or magnetic resonance imaging. Instead, we planned to perform a staging total body TC (or PET-TC) as soon as a histological examination was available.

The patient signed an informed consent to surgery and underwent surgery under local anesthesia in the outpatient care.

After cutaneous incision, the tumor appeared as a whitish mass, wooden-fibrous in consistency, strictly adhering to the skin, and apparently fixed to the deep planes. Carefully avoiding

tumor rupture, the tumor was excised after identification of the cleavage plane between the surrounding layers and the mass, both using a swab mounted on a clamp for blunt dissection and using scissors (Figure 1). After careful hemostasis by electrocoagulation, the wound was sutured. The postoperative course was uneventful, and the patient was discharged the same day.

Upon microscopic examination, a sclerosing perineurioma was reported. Neoplastic cells were immunoreactive for CD34, CD99 (Figure 2), and EMA (Figure 3) and negative for S100 protein, smooth-muscle actin, pancytokeratin (AE1–AE3), CD31, neurofilaments, and beta-catenin, respectively.

According to the benign biological tumor behavior, radical resection was considered an adequate treatment, and further radiological staging exams were not performed. The patient had no signs of relapse after 24 months of follow up.

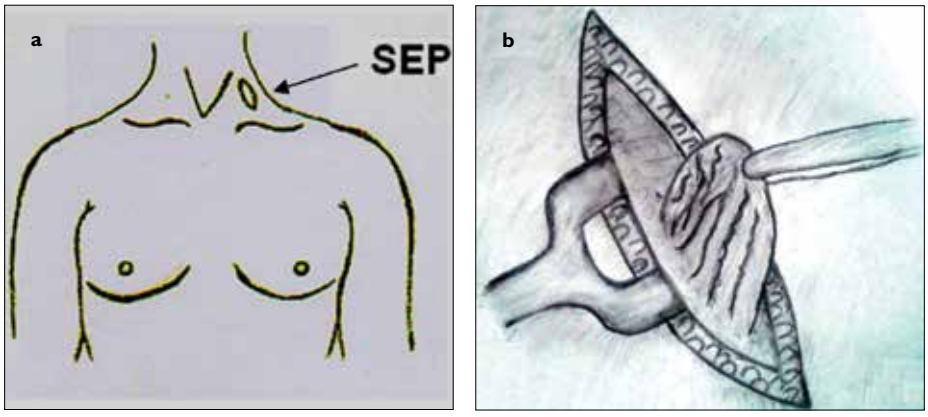

Figure 1. a, b. Clinical presentation drawing (a) shows the left supraclavicular swelling of approximately 1 cm (indicated by the arrow), resulted to be a SEP at histological examination. Intraoperative drawing (b) shows a whitish mass, wooden-fibrous in consistency, strictly adhering to the skin and apparently fixed to the deep planes

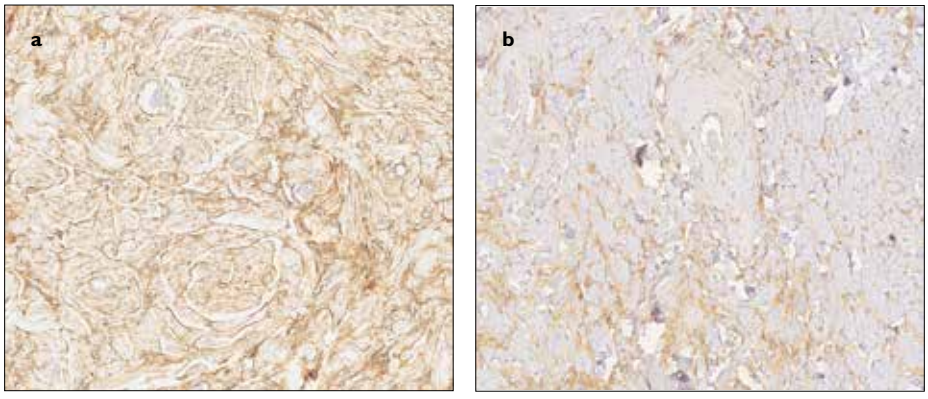

Figure 2. a, b. Microscopic view at immunohistochemical study shows CD34 (a) and CD99 (b) positivity

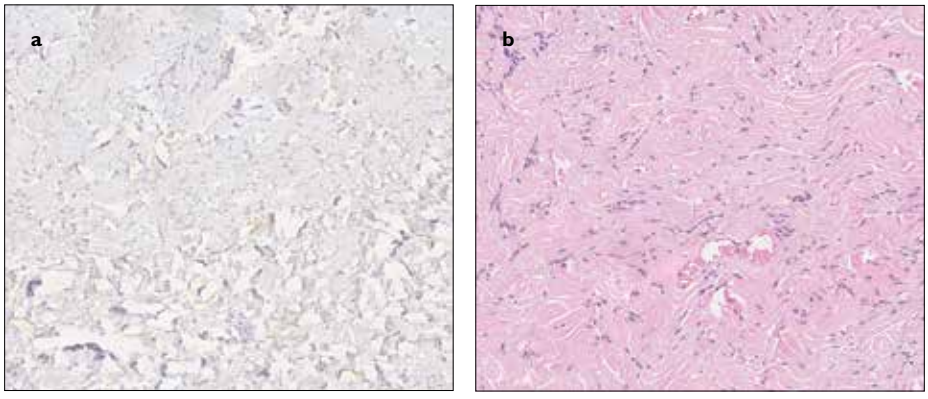

Figure 3. a, b. Microscopic view at immunohistochemical study (a) shows EMA positivity (epithelial membrane antigen) and microscopic view at histological study (b) shows a hematoxylin-eosin staining

Discussion

The sclerosing subtype of extraneural perineurioma (SEP) was first described in 1997 by Fetsch and Miettinen, and approximately 60 cases have been reported to date in medical literature [6-8]. SEP characteristically occurs on the hands as a single asymptomatic fibrous papule or nodule, and only few cases of non-acral or multiple presentations have been described [1, 3]. It is equally common in males and females with peak incidence in the middle age, although a dozen cases were also reported in pediatric age [1].

Histologically, the SEP is a peripheral nerve sheath tumor originating from the perineural cells and presenting as a well-circumscribed and hypocellular nodule located in the dermis or hypodermis [1-3]. The small epithelioid and spindle cells are arranged in an onion bulb-like and trabecular growth pattern within a dense collagen stroma [1-3]. The thin elongated and bipolar cytoplasm is mostly arranged in a storiform and whorled architecture, whereas the nucleus is tapering and presents a low mitotic activity [1-3].

Since the morphological features of the SEP are shared with other tumor types, the histopathologic diagnosis often becomes difficult, and immunohistochemistry studies become mandatory. The perineural differentiation is immunohistochemically demonstrated by EMA positivity and S100 protein negativity [2, 3]. Additionally, medical literature reported cases where the SEP was immunopositive for collagen type IV, laminin, vimentin, claudin A, cytokeratins, CAM 5.2, muscle-specific actin, GLUT-1, CD31, CD34, and CD99 [2, 3].

When immunohistochemistry is unable to differentiate the SEP from other tumors, the detection of some cytogenetic abnormalities may be useful for diagnosis; for example, low-grade fibromyxoid sarcoma may present EMA positivity, and the finding of a translocation involving 16p11.2 allows its diagnosis [2]. More frequently, differential diagnosis must be conducted with tendon sheath fibroma, sclerotic fibroma associated with Cowden syndrome, epithelioid neurofibroma, giant cell tumor of tendon sheath, and fibrosing adnexal tumors [3]. Some authors consider the perineurioma to be a peripheral form of meningioma, and the association of SEP and neurofibromatosis is rarely described [2]. The behavior of SEP is benign, although rare cases of malignant perineurioma have been reported [1-9].

Imaging studies, such as computed tomography and magnetic resonance imaging, may help define the surgical approach in case of a particular location, such as visceral or retroperitoneal, but the definitive diagnosis is always immunohistochemical and/or cytogenetical. Surgical excision is the only and definitive treatment [1-9].

The present case is the starting point for some considerations. First, our case has an unusual non-acral location and is the first SEP reported in medical literature with clinical presentation as supraclavicular swelling mimicking a lymph node tumor. Second, as discussed, perineural differentiation is immunohistochemically demonstrated by EMA positivity and S100 protein negativity. Instead, few cases have been also reported with immunopositivity for CD34 and CD99 [2, 3]. Our case also presents positivity for CD34 and CD99 (Figure 2). Third, we did not perform preoperative radiological examination of the mass because we have had trust in clinical presentations that had led us to suspect a lymph node neck tumor. We take this opportunity to reiterate that, in such cases, it is always important to perform at least a preoperative ultrasonographic imaging, which, although not changing the medical approach (i.e., removal for histological examination and diagnosis), allows to have more preoperative anatomical information about the mass, and therefore to be better prepared for the operation.

Fourth, given its rarity, we consider important to spread and share our experience with SEP because clinicians should be aware of its occurrence and keep it in mind during the differential diagnostic process.

Informed Consent: Written informed consent was obtained from patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - E.I.; Design - A.T.; Supervision - M.T.; Resources - P.G., L.D.; Materials - P.G.; Data Collection and/or Processing - L.D., E.I.; Analysis and/or Interpretation - M.T.; Literature Search - A.T.; Writing Manuscript - A.T., P.G.; Critical Review - M.T., P.G.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study has received no financial support.

References

1. Bhat W, Akhtar S, Teoh V, Bourke G. Sclerosing Perineuroma in paediatric fingers: A rare distinct and underrecognised entity. J Plast Reconstr Aesthet Surg 2014; 67: 1161-2. [CrossRef]
2. Duff DJ, Guzman MA, Batanian JR. ABL1 gene involvement within a complex three-way translocation (2;9;4) in perineurioma characterized by molecular cytogenetic methods. Cancer Genet 2014; 207: 263-7. [CrossRef]
3. Armengot-Carbo M, Millan F, Sanjuan J, Quecedo E, Gimeno E. Sclerosing perineurioma: case report and literature review. Dermatol Online J 2014; 20.
4. Lazarus S, Trombetta L. Ultrasound identification of a benign perineural cell tumour. Cancer 1978; 41: 1823-9. [CrossRef]
5. León Cejas L, Binaghi D, Socolovsky M, et al. Intraneural perineuriomas: diagnostic value of magnetic resonance neurography. J Peripher Nerv Syst 2017. [CrossRef]
6. Fetsch JF, Miettinen M. Sclerosing perineurioma: a clinicopathologic study of 19 cases of a distinctive soft tissue lesion with a predilection for the fingers and palms of young adults. Am J Surg Pathol 1997; 21: 1433-42. [CrossRef]
7. Toussaint-Caire S, Aguilar-Donis A, Torres-Guerrero E, et al. Sclerosing acral skin perineurioma: clinicopathologic study of ten cases (eight classical and two with xanthomatous changes). Gac Med Mex 2015; 151: 299-305.
8. Kaku Y, Fukumoto T, Louise Opada G, Anan T, Kimura T. A clinicopathologic and immunohistochemical study of 7 cases of sclerosing perineurioma. Am J Dermatopathol 2015; 37: 122-8. [CrossRef]
9. Vajtai I, Hewer E, Andres R, Neuenschwander M, Kappeler A, Gugger M. Meningial perineurioma: a benign peripheral nerve sheath tumour in a previously unrecognized central nervous system location, mimicking meningioma. Pathol Res Pract 2011; 207: 592-6. [CrossRef]