In Reply to the “Letter to the Editor Regarding Intraneural Ewing Sarcoma of the Fibular Nerve—Case Report, Radiologic Findings and Review of Literature”

We read with interest the reply to our article written by Boffano and Piana. Their interest in the topic allows us to further deepen the details of the presented case report and to highlight the current role of biopsy in peripheral nerve tumors. Nevertheless, restating the correct management of soft tissues sarcomas (STS) was not the aim of our article.

First of all, the matter at issue was not a common STS but rather an intraneural Ewing sarcoma (ES), which is an unquestionably rarer pathologic entity. We believe that considering it as an STS with a rare onset site could be incorrect and approximate from clinical, diagnostic, and surgical points of view. Although, for STS, recent and complete clinical practice guidelines have recently been published to help in the management of these tumors, for the extraosseous ES and in particular for intraneural ES, little is mentioned.

In our case, the ultrasound findings were consistent with a benign peripheral nerve sheath lesion. Magnetic resonance preoperative findings were also consistent with the typical aspect of a benign peripheral nerve sheath tumor, even if some sequences could not allow an unequivocal interpretation.

For this reason, we decided to perform a marginal excisional biopsy for a histologic verification, as recommended by other authors in similar cases. During surgery the suspicion of a malignant tumor was more consistent, due to the appearance and macroscopic aspects of the lesion that differed from the typical benign nerve sheath tumors.

Indeed, incisional and excisional biopsies are essential to get an adequate therapeutic management, as suggested by current clinical practice guidelines and, moreover, allow surgeons to perform more radical second surgery interventions, if necessary. Nerve biopsy strategy is quite different from soft tissues biopsy in terms of feasibility and postprocedural risks and complications. Needle or Tru-Cut biopsies in peripheral nerve lesions could be inaccurate in similar cases due to the minimal sampling that they can supply and for the consistent risk of intense neuropathic pain. Nevertheless, they could be considered when there is a suspicion on radiologic findings, clinical examination, and behavior features of the tumor, elements that were not present in our clinical case.

Although clinical practice guidelines do exist for a correct management of the patient and preoperative decisions about the treatment of STS, less is known about malignant peripheral nerve sheath tumor management, especially when these tumors clinically and radiologically mimic benign peripheral nerve sheath lesions. Also, almost nothing is known about the management of such specific cases like intraneural ES. When there are no clinical and radiologic elements to suspect a malignant lesion of a peripheral nerve, a preoperative multidisciplinary setting is usually not gathered, so intraoperative and postoperative management becomes more challenging.

Being aware of the possibility that such rare malignant pathologies of peripheral nerves are similar to more frequently observed benign tumors could contribute to better define the management of these oncologic entities in the future.

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REFERENCES