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CASE REPORT

Successful Surgery Management of Giant Periorbital Malignant Schwannoma in a Low-Income Setting

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Abstract Schwannoma can arise from any cranial, peripheral or autonomic nerve, except the olfactory and optic. About 25–45% of extracranial schwannomas lie in the head and neck. Data on malignant schwannoma from low-income settings are inconsistent. We reported a case of giant periorbital malignant schwannoma successfully treated in a low-income setting. The strength of our case is given not only by the rarity and the size of the disease but also for highlighting the weakness of health system in low-resource settings. It is mandatory to strengthen the health system with particular attention to physical, psychologic and social aspects and to promote comprehensive programs including all these aspects.

Keywords Schwannoma · Low-income settings · Head tumor · Malignant schwannoma

Introduction

Tumors involving the peripheral nervous system can be intrinsic or extrinsic, and both can be either benign or malignant [1]. Benign peripheral nerves sheath tumors include schwannomas (or neurilemmomas or neurinomas) and neurofibromas. Most cases are sporadic, but can also be associated with neurofibromatosis or schwannomatosis [1].

Schwannoma can arise from any cranial, peripheral or autonomic nerve, except the olfactory and optic. About 25–45% of extracranial schwannomas lie in the head and neck [2]. There are four major forms of schwannoma: conventional, cellular, plexiform and melanotic. Malignant change in schwannomas of the head and neck is rare, with a prevalence of 8–13.9% [2].

Malignant peripheral nerve sheath tumors show variable differentiation toward one of the cellular components of nerve sheaths (Schwann cells, fibroblasts or perineurial cells) [3]. They account for 5% to 10% of soft tissue sarcomas and have an incidence of 0.001% in the general population. They can occur sporadically, as well as in patients with neurofibromatosis, and arise either de novo or from a preexisting benign tumor [3].

Malignant peripheral nerve sheath tumor occurs most frequently in the extremities, particularly proximally, followed by the trunk and head and neck [1]. Patients may present with a painful or rapidly enlarging mass with associated neurologic deficits. Poorer prognosis is associated with large tumors (with size varying from > 5 to > 7 cm in different studies) and incomplete resection [1]. Radical surgery is the mainstay of current management, while radiotherapy may be used to control local disease and reduce recurrence, but it has little effect on long-term survival [3]. Chemotherapy is generally not effective, although some studies have shown that it may benefit patients with high-grade histology or children with unresectable tumors [3]. Data on malignant schwannoma from low-income settings are inconsistent. We reported a case of giant periorbital malignant schwannoma successfully treated in a low-income setting.

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Case Presentation, Management, and Outcome

A 20-year-old man presented in Beira Central Hospital, Mozambique, with a giant mass located in the zygomatic and frontal region evolved over about 1 year (Fig. 1a). The only symptoms were mild pain and increasing visual disturbance due to the looming mass. He disclosed no significant past medical history, general condition was good, and he had normal vital parameters. The computed tomography (TC) showed a well-defined soft tissue mass (Fig. 2a, b). The differential diagnosis included myxoma, neurofibroma and dermoid cyst.

He was HIV negative, and preoperative tests showed normal parameters. The patient underwent surgical excision, and we enucleated the mass measuring 13 × 8 × 5 cm (Fig. 1b). Histologic examination showed Schwann cells in a collagenous matrix, and the diagnosis was a malignant schwannoma. The post-surgical course was regular, and the 10-day follow-up showed a clean scar (Fig. 3a, b) and no sequela.

Discussion

Malignant schwannomas arising in the peripheral nerves can develop every anatomic region although those of the head and neck are quite rare [4]. We reported a late-stage giant malignant schwannoma in low-income countries. There are at different reasons that could explain this advanced presentation, as in our case only after 1 year and due to a heavy visual impairment. First of all, in low-income countries, traditional healers play a major role in society, especially in rural areas. This has worsened many times by the lack of highly qualified or specialized health workers and, thus, by the lack of trust in conventional services. Again, distance from the hospital or health center represents in many cases a socioeconomic barrier to healthcare access. When a patient finally turns to the health system, his health path faces many challenges. In fact, although a multidisciplinary and tailored approach is suggested especially for oncologic patients, in low-resource settings it is not possible both due to the lack of specialized health workers and the absence of adequate equipment and drugs for diagnosis, therapy and follow-up. In our case, we were unable to have a bioptic confirmation of the diagnosis before the surgery, although our work was facilitated by the TC. The surgery was performed by a general surgeon because in the hospital there is no plastic surgeon and, considering the mass localization, it could represent a reason for social discrimination. Finally, although the surgery is the gold standard, there is no oncologist nor chemo and radiotherapy and this is a great weakness for a health system even more so if we consider that our is the second and referral hospital in the country.

The strength of our case is given not only by the rarity and the size of the disease but also for highlighting the weakness of the health system in low-resource settings. It is mandatory to strengthen the health system with particular

Fig. 1 Giant schwannoma at presentation (**a**) and the excised mass (**b**)

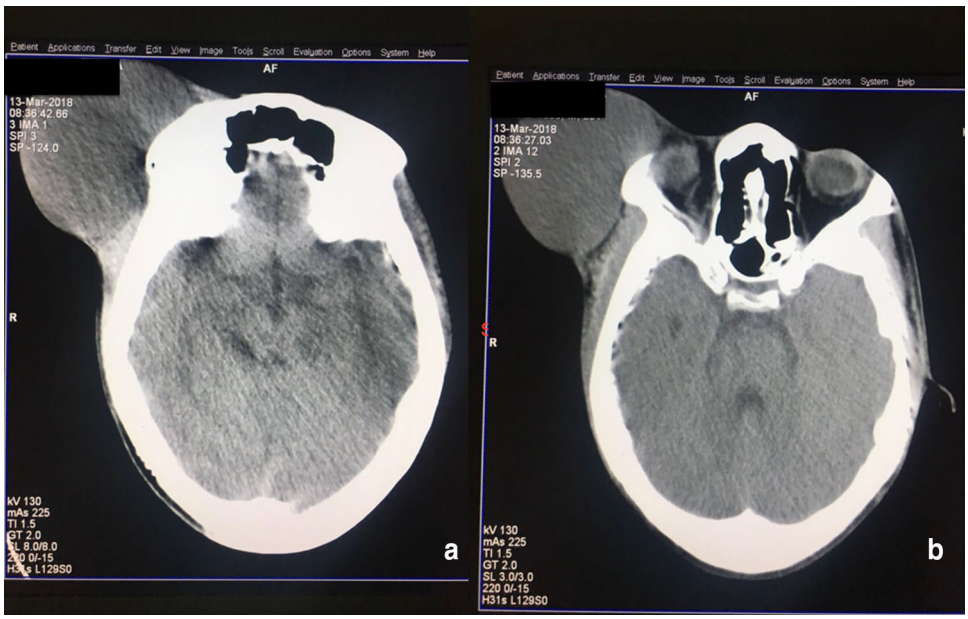

Fig. 2 Computed tomography showing the schwannoma in the periorbital region (**a** and **b**)

Fig. 3 The 10 days post-surgical presentation of clean scar (**a** and **b**)

attention to physical, psychologic and social aspects and to promote comprehensive programs including all these aspects.

Compliance with Ethical Standards

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

Informed Consent Written informed consent was obtained from the patients for publication of this case report and any accompanying images.

References

1. Montano N, D'Alessandris QG, D'Ercole M, Lauretti L, Pallini R, Di Bonaventura R, La Rocca G, Bianchi F, Fernandez E (2016) Tumors of the peripheral nervous system: analysis of prognostic

factors in a series with long-term follow-up and review of the literature. J Neurosurg 125(2):363–371
2. Agrawal A, Singh V, Bhagol A, Kumar P, Narwal A (2017) Extracranial trigeminal schwannomas: a retrospective analysis. J Maxillofac Oral Surg 16(2):164–169
3. Behuria S, Rout TK, Pattanayak S (2015) Diagnosis and management of schwannomas originating from the cervical vagus nerve. Ann R Coll Surg Engl 97(2):92–97
4. Das Gupta TK, Brasfield RD (1970) Solitary malignant schwannoma. Ann Surg 171(3):419–428