European Annals of Otorhinolaryngology, Head and Neck diseases 133 (2016) 23–26

Available online at ScienceDirect www.sciencedirect.com

Elsevier Masson France EM|consulte www.em-consulte.com/en

Original article

Unusual outer ear swelling: Childhood auricular rhabdomyosarcoma

M. Chirat[a], L. Dainese[b], S. Fasola[c], V. Couloigner[a], F. Denoyelle[a], E.-N. Garabedian[a], N. Leboulanger[a,*]

[a] Service d'ORL et de chirurgie cervico-faciale, hôpital universitaire Necker Enfants Malades, Paris V University, 149, rue de Sèvres, 75015 Paris, France
[b] Service d'anatomo-pathologie, hôpital Armand-Trousseau, Paris VI University, 26, avenue du Dr Arnold-Netter, 75012 Paris, France
[c] Service d'onco-hématologie, hôpital Armand-Trousseau, Paris VI University, 26, avenue du Dr Arnold-Netter, 75012 Paris, France

ARTICLE INFO

Keywords:
Rhabdomyosarcoma
Outer ear
Auricle
Child

ABSTRACT

Introduction: Rhabdomyosarcoma (RMS) is the most frequent soft-tissue sarcoma in children and makes up 5% of all pediatric malignant tumors. The main head and neck locations are the base of the skull, nasopharynx, nasal cavity and orbit. An outer ear location is considered extremely rare. We present 3 cases of children, aged 6 to 14 years, presenting with auricular RMS.
Case reports: The first child, aged 6, was managed by 4 chemotherapy cycles followed by surgical resection of the tumor bed, completed by 5 further cycles of chemotherapy. The second, aged 14, was managed by 4 chemotherapy cycles followed by external radiation therapy of the tumor bed and lymph node areas, completed by 5 further cycles of chemotherapy. The third, aged 13, was managed by 4 chemotherapy cycles followed by surgery, completed by 5 further cycles of chemotherapy.
Discussion: In these 3 patients, the treatment program achieved complete disease control. Prognosis was good, thanks to good surgical access. Diagnosis should be considered in case of unusual progressive swelling in the outer ear.

1. Introduction

Rhabdomyosarcoma (RMS) is the most frequent soft-tissue sarcoma in children and makes up 5% of all pediatric malignant tumors [1]. Onset is usually before the age of 12 years [2]. In pediatric populations, 2 main histologic forms are seen: embryonal and alveolar RMS. Pleomorphic RMS is mainly found in adults.

In children, 30% to 40% of RMSs develop in the head and neck region, the main locations being the base of the skull, nasopharynx, nasal cavity and orbit [3,4]. RMS is a malignant mesenchymatous tumor growing from primitive cells involved in skeletal muscle differentiation. It may also occur throughout the body, even in organs without striate muscle [1,3].

Outer ear locations are considered extremely rare; to the best of our knowledge, there have been only 3 case reports [4–6]. Rarity, absence of specific symptoms and misleading, sometimes inflammatory and infectious presentation can delay diagnosis and lead to unsuitable primary management. We report 3 new cases managed by a single team, and discuss treatment modalities according to existing international guidelines.

2. Material and method

A retrospective analysis was performed on files of patients followed for auricular RMS by the same team in the pediatric ENT departments of the Necker Hospital for Sick Children and Armand-Trousseau Hospital, Paris, France, over a 4-year period from November 2010 to November 2014.

3. Results

Three children presenting with auricular RMS were treated during the study period.

3.1. Patient #1

A 6-year-old boy presented with swelling of the left concha of 5 months' evolution. The lesion resembled a cheloid scar, but there was no history of trauma or infection. Given the persistence and growth of the lesion (2 cm long axis at diagnosis) (Fig. 1), biopsy was performed under general anesthesia.

Microscopy found skin cover with islands and clusters of malignant tumoral proliferation. The tumor cells were mostly round, with clear cytoplasm and a nucleolated fine chromatin nucleus. There were numerous mitoses. There were also, more rarely,

* Corresponding author.
E-mail address: nicolas.leboulanger@nck.aphp.fr (N. Leboulanger).

http://dx.doi.org/10.1016/j.anorl.2015.09.005
1879-7296/

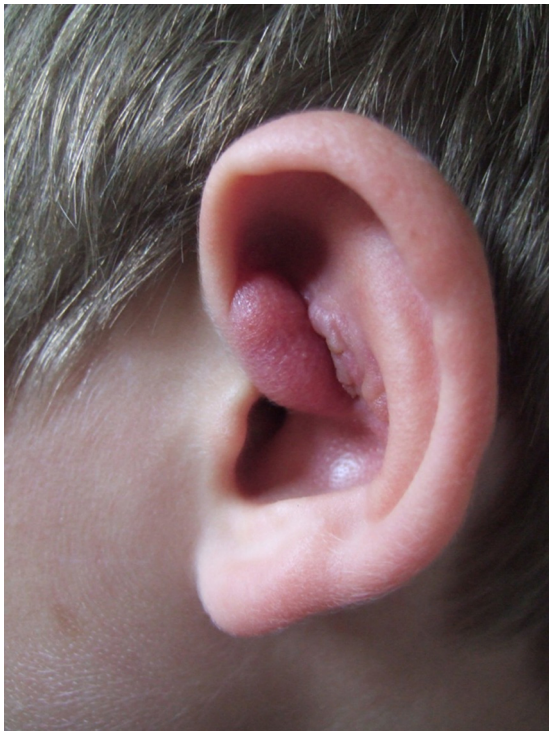

Fig. 1. Patient #1, primary lesion at biopsy.

cells with ribbon- or racket-shaped eosinophil cytoplasm. On immunohistochemistry, about 35% of tumor cells were desmin and myogenin positive (Fig. 2a–c). RT-PCR screening for specific solid tumor translocation was negative. Embryonal RMS was diagnosed.

Extension assessment comprised brain and temporal MRI, head and neck and thoracic contrast-enhanced CT, bone scintigraphy and myelogram. In the left auricle, MRI found an a bumpy contoured oval mass 26 mm high, 13 mm wide and 20 mm thick, showing weak signal on T2 and FLAIR sequences, with strong uptake and relatively well-delineated. Cerebral parenchyma was normal. CT showed the formation of the outer ear soft tissue, without bone extension. There was no cervical adenomegaly or metastatic extension on chest CT or bone scintigraphy.

There was thus N–, IRS III localized embryonal RMS of less than 5 cm in a favorable location in a child less than 10 years of age.

The multidisciplinary team meeting recommended implementing the EpSSG RMS 2005 standard risk group, subgroup C protocol [7]. The patient therefore initially received 4 cycles of IVA (ifosfamide, vincristine, actinomycin); clinical and radiological response was good after 3 cycles, and surgical resection of the tumor bed was performed.

Microscopic inspection of the specimen found no viable residual tumor; all margins were healthy.

Resection being macro- and microscopically complete, 5 postoperative cycles of IVA were implemented, without radiation therapy. At the time of writing (36 months' follow-up), the patient was in complete remission. Secondary auricleplasty may later be considered, depending on the patient's esthetic desiderata.

3.2. Patient #2

A 14-year-old boy presented with a lesion of the left concha. It was initially associated with juvenile acne and later attributed to a sebaceous cyst. Several antibiotic and local treatments were without effect; surgical resection was also suggested. The swelling progressively increased over 7 months, and the parents consulted a hospital dermatology department. Examination found a large

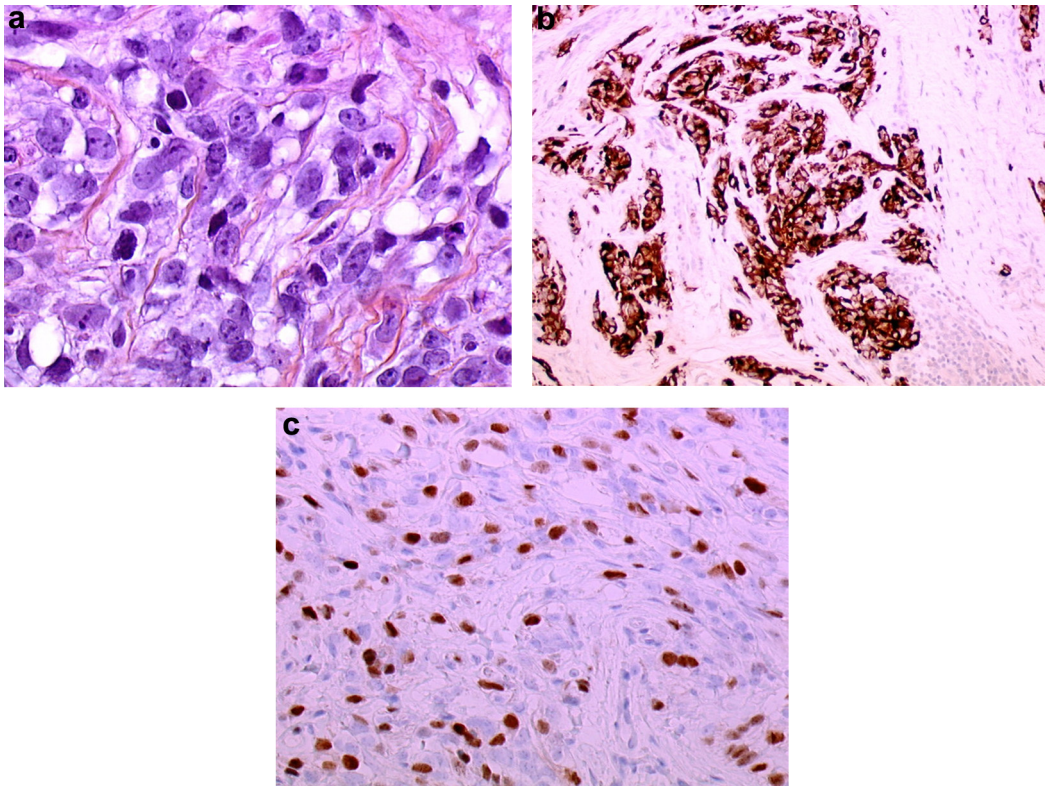

Fig. 2. a: patient #1, malignant proliferation comprising islands separated by thin fibrous septa. Cells of various sizes, with poorly visible cytoplasm, round or oval nuclei with jutting nucleoli. Frequent mitoses. Hematoxylin-eosin staining, × 40 magnification; b: patient #1, desmin staining, × 20 magnification: intense membrane and cytoplasm staining and diffuse tumor cell staining; c: patient #1, myogenin staining, × 20 magnification: positive tumor cell nuclei, assessed at 35%.