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CASE STUDY

Rhabdomyosarcoma of the Nasal Vestibule in a Child

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السَّاركومةُ العَضَلِيَّةُ المُخَطَّطَة في دِهْليزُ أَنْف طفل

الملخص: نقدم حالة السَّاركومةُ العَضَلِيَّةُ المُخَطَّطَة في طفلة بعمر سنتين بدون وجود عامل مُؤَهِّب مسبق لحصوله . لا تؤشر حالتهاالسريرية على تشخيص المرض الذي جاء كمفاجأة من المختبر . كان تقرير الفحص النسيجي المرضي هو السَّاركومةُ العَضَلِيَّةُ المُخَطَّطَة السِنْخِيّة.

المفردات المفتاحية: الساركومة العضلية المخططة الانفية . الطفولة . تقرير حالة . عمان .

ABSTRACT A case of rhabdomyosarcoma in a 2 year old girl without a pre-existing predisposing factor visited the ENT Department of Sultan Qaboos University Hospital (SQUH). Her clinical condition did not point to the diagnosis, which came as a histological surprise. The histopathology report was alveolar rhabdomyosarcoma.

Keywords: Nasal rhabdomyosarcoma, pediatric age, Case Report, Oman.

About 5-8% of childhood tumours are Rhabdomyosarcomas (RM).[1] A sarcoma is a malignancy originating from primitive mesenchymal cells, which under normal circumstances develops in supportive tissues such as muscle and bone.

THE CASE

A two year old girl was brought in the Ear Nose Throat (ENT) Department of SQUH by her parents with a mass blocking the right anterior nares completely, causing difficulty in breathing. The blockage had lasted already several weeks. There were no other symptoms like itching or bleeding.

On examination, the child looked healthy, but had a spherical mass about 1cm x 1cm in diameter attached to the skin of the floor of the nasal vestibule. This was covered with intact skin, which although it looked pink had no ulceration or tenderness. The mass was not fixed to the surrounding tissue. There were no palpable cervical glands. The left nasal cavity and the oral cavity were normal. On admission and after a complete blood count and blood chemistry all results were found to be normal.

It was obvious that an excision biopsy would provide the correct diagnosis. This was done under general anesthesia with an oro-tracheal intubation and a pharyngeal pack. The swelling was actually shaved out of the nasal vestibule taking care not to injure the cartilage of the vestibule. The bleeding was very mild and quickly stopped by application of pressure and a small anterior nasal pack. The specimen was sent to the histopathology department.

Postoperatively, the child made an uneventful recovery. The nasal pack was removed after 24 hours and the denuded area of the vestibule start to heal well without problems. The area was painted daily with antiseptic cream and the child was given prophylactic antibiotic for seven days (Augmentin syrup). The specimen was sent to the histopathological department and the result came as RM of the alveolar type with positive monoclonal antibodies to desmin (tumour related antigen).

The child was referred to the pediatric oncologist and was prescribed chemotherapy. It is well known that RM is chemosensitive, with the use of multiple drugs [(5)] normally required. The intensity of treatment depends on

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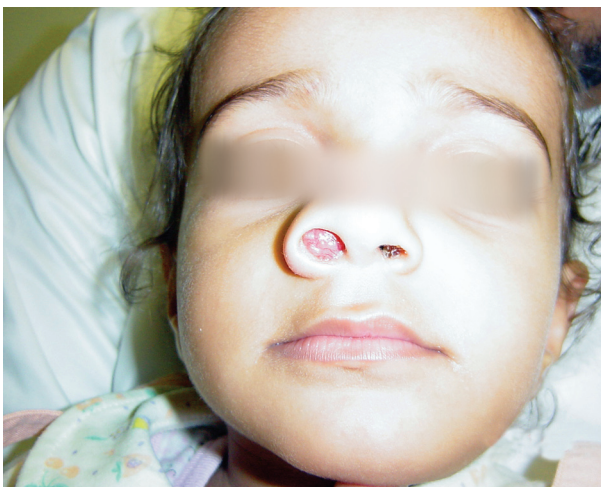

- The stage of the sarcoma
- The site
- The histopathology

The child initially recovered well, however she did not report one month later for the usual follow up.

DISCUSSION

Most sarcomas show some evidence of differentiation into a particular supportive tissue, but maturation is incomplete.

The most common normal tissue counterpart for childhood sarcomas is skeletal muscle, and tumors having features of skeletal muscle differentiation are called rhabdomyosarcomas.

A small subset of children with rhabdomyosarcomas, mostly genito-urinary rhabdomyosarcomas, belong to families in which multiple cancers such as breast lung and brain tumors occur. Adrenocortical cancer and leukemia can also develop. Constitutional mutation in the p53 can be found. The tumor suppressor gene located on the long arm of chromosome 17, characterizes this cancer predisposition syndrome (the Li Fraumeni Syndrome).

RM arise from mesenchymal tissue. The head and neck region is the most common site of origin in 40% of cases, the genito-urinary in 20% of cases, 20% in the extremities and 10% in the trunk.[6]

RM is the most common type of head and neck cancer in the first 2 years of life.[1] 50% of head and neck tumors arise in the orbit and the nasopharynx.

SUBDIVISION

25% arise in the orbit, 50% in non-orbital parameningeal sites (the nasopharynx, middle ear, mastoid and pterygoid and infra temporal fossa) 25% are non-orbital non-parameningeal (the mouth, the neck, face, scalp, larynx).[2]

Symptomatology

It usually presents as a painless mass. In the larynx it results in upper airway obstruction. In a few cases there is persistent pain with sanguinous discharge and cranial nerve palsies. In the areas covered with mucous membrane it can have a botryoid (bunch of grapes) appearance.

Diagnosis

This can be done by MRI scan set at bone windows and/or biopsy.

Histopathology

Spindle shaped cells with eosinophlic cytoplasm or cross striation like striated muscles. However, the most common appearance is that of an undifferentiated small, round, blue cell tumour with the differential diagnosis of neuroblastoma, Ewing's sarcoma, primitive neuroectodermal tumour or non Hodgkin's lymphoma. Monoclonal antibodies which are directed against the tumor-related antigens, such as desmin in RM or leucocytes common antigen (in case of lymphoma) or neuron specific enolase (in the case of neuroblastoma or primitive neuroectodermal tumors), can help in a differential diagnosis.

Pathological sub-types

1. Embryonal (botryoid) variant with good prognosis, arising in a sub-mucosal location.
2. Alveolar with poor prognosis, due to aggressive behaviour and tendency to metastasize early.
3. Pleomorphic, the most common in adults, rare in children

Spread

The tumor spreads to regional lymph nodes in 5% of cases and to lung, liver, bone and bone marrow, with 4 stages of tumor according to the degree of spread.

Treatment

1. Surgical complete excision or biopsy. However, successful complete excision (biopsy) is normally achieved in only 4 out of 60 patients.
2. Multiagent chemotherapy, judicious surgery and radiotherapy to selected patients
3. Chemotherapy (chemo sensitive) with cyclophosphamide and ifosfamide.

Actinomycin D, vincristine, Doxorubicin C, Toposide, platinum analogues like cis and carboplatinum. At least 3 drugs should be given.

Prognosis

1. 5 years survival: 92% with in orbital RM.
2. 81% with non-parameningeal non-orbital.
3. 69% with parameningeal.

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