CASE STUDY

Tufted Angioma of the Face

Angioma en penacho de la región facial

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Clinical Case

We present the case of a 9-year-old female patient that was sent to our service because of a purplish lesion on the bridge of the nose. This lesion was seen at the age of 7 as a small, asymptomatic red wine macula, which grew progressively. At present it is slightly painful and presents popular areas.

Physical examination revealed that there was a 2 cm × 1 cm purplish oval plaque, indurated and slightly adhered to the deep planes. The lesion presented 2 nodular areas in the underlying skin in both nasal bones (Fig. 1). The rest of the skin and the ENT examination of the ENT area was normal. The blood counts and biochemical blood analyses were within the established biological ranges.

Faced with the suspicion of in-depth infiltration in the area of the nasal bones, we ordered a computed tomography. This test ruled out signs of bone and orbit infiltration.

Given the location of the lesion, it was surgically removed. The lesion described was resected up to the deep bone plane. After the excision the resulting defect was reconstructed using a glabellar flap (Fig. 2).

Figure 1 Preoperative image. Purplish lesions on the bridge of the nose.

Pathology analysis of the surgical specimen yielded the definitive diagnosis of tufted angioma (TA). The lesion corresponded to a neoplasia of endothelial cells arranged in lobules, with irregularly rounded or ovoid nuclei, a small nucleoli and clear eosinophilic cytoplasm with poorly defined borders (Fig. 3). The index of proliferation was low (Ki67 lower than 1%). The patient came to our service for periodic follow-up and there is currently no evidence of recurrence.

Discussion

The TA is a histologically benign tumour of vascular origin. The name TA was first used to describe these tumours by Wilson Jones in 1976. However, these lesions had already
been described half a century earlier (1949) by Japanese authors that used the name of angioblastoma of Nakagawa. In 1971 similar lesions were described as progressive capillary haemangiomas. It is currently postulated that all these descriptions represent the same lesion.

The prevalence of TA is unknown, with there having been some 200 cases described to date. Histologically, TA is characterised by capillary lobules arranged in the dermis with a ‘buckshot’ distribution. It normally appears in the first 5 years of life, but a fourth of the cases present during adulthood. It is most frequently found on the trunk and lower limbs and very rarely on the face, although some cases have been described in the cervical area. Clinically it presents as a subcutaneous, red-wine plaque or nodule. When palpated, the lesion can be slightly painful. It usually presents with a border more indurated and having a central depression, so it has been compared to the shape of a ‘donut’, located subcutaneously. Typically, these lesions slowly grow laterally over various months or years.

In differential diagnosis of TA, kaposiform haemangioendothelioma (KHE) should be considered. A tumour of characteristics similar to TA, KHE originates in the spiny endothelial cells, which present in the paediatric ages and are locally aggressive; although it is benign histologically, it can infiltrate to great depths. In contrast to the common haemangiomas more frequent in girls, KHE occurs just as often in both sexes. Recently, the expression of marker D2-40 has been shown in these 2 lesions, but not in other haemangiomas, which suggests that these tumours can be part of the spectrum of the same condition. In initial stages it can be confused with a haemangioma, but as it progresses its pattern of growth is nodular with the aspect of a malignant tumour. Both KHE and TA can be associated with Kasabach–Merritt syndrome. The remission of these lesions

Figure 2 (A) Skin defect caused by the tumour removal. (B) Postoperative result, at 7 days.

Figure 3 Pathology image of tufted angioma. (A) Lobules of small capillaries scattered over the dermis in a ‘buckshot’ pattern (haematoxylin-eosin, original magnification 20×). (B) Each lobule is composed of aggregates of endothelial cells lacking atypia or mitosis, which surround capillary lumina that can contain erythrocytes (arrow), (haematoxylin-eosin stain, original magnification 200×).
is rare, although it has been described in periods ranging from 5 months to 2 years.\(^5\)

The most appropriate mode of treatment for these lesions depends on the characteristics of the lesion on its placement. In our case, the lesion was expanding as it grew and it was located in the centre of the face, near the eye area. Consequently, we chose radical excision of the lesion to prevent, if the medical treatment was ineffective, the lesion from reaching critical areas.

Various options exist for medical treatment. The use of systemic or intralesional corticoids, low doses of acetylsalicylic acid, IFN-alpha and even radiotherapy and chemotherapy with vincristine and pulsed laser have shown varying results.\(^7\) The use of propranolol as the treatment of choice in common infantile haemangioma has recently been approved. However, propranolol is not indicated in the case of TA because it is a histopathological entity and has a different natural history.\(^8\)

When the lesions are small or in critical areas (such as in our case), surgical removal and posterior reconstruction is the treatment of choice.\(^9\) The proximity of structures that can lead to important functional deficits if they are invaded, as well as the no less important aesthetic defect that they can cause, make surgical planning even more complex. That is why individualisation of treatment is essential in these cases. We decided to use a glabellar flap to reconstruct the area as the tumour invaded soft tissues and perios-teum deeply, as a free graft could be more likely to develop necrosis.

**Conclusion**

Although infrequent, TA should be borne in mind in differential diagnosis of vascular lesions in the craniofacial area. The marker D2-40 has been found expressed in both TA and KHE, which suggest that these tumours may be part of the spectrum of the same condition. At present, there is no consensus on the treatment of this type of tumour. In the case of lesions that are small or in those located in areas that are critical due to their proximity to important structures, surgical treatment provides good results, both functionally as well as aesthetically.

**Conflicts of Interest**

The authors have no conflicts of interest to declare.

**References**