

An Unusual Mole: An Adult Case of Dabska Tumour

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ABSTRACT

In 1969 Dabska and her colleagues described for the first time this rare malignant tumour, also later known as a malignant endovascular papillary angioendothelioma of childhood. Overall, depending amongst other factors on its location, it is thought to have a generally favourable prognosis and a wide local excision seems to be the treatment of choice. We here present a very rare and unusual case of a 63 year old woman with a 20 year history of slow-growing right buccal dermatological lesion which resembled a common mole. The histopathological diagnosis of Dabska Tumour was made following the hematoxylin and eosin (H&E) biopsy. The analysis revealed multiple delicate interconnecting vascular channels with papillary plugs, some of which containing hyalinized core, projecting into the lumen lined by atypical plumped endothelial cells.

Key words: Dabska Tumour, angiosarcoma, histopathology

Introduction

The Dabska Tumour (DT) is a very rare low-grade angiosarcoma with a generally favourable prognosis for which a wide local excision seems to be the treatment of choice¹. It is also known as a malignant endovascular papillary angioendothelioma of childhood and at low power microscopy there is a resemblance to a cavernous lymphangioma¹. Since its first description in 1969 by Dabska et al. only about 30, predominantly paediatric, cases have been described^{1–6}. Recently, a paediatric occurrence within a congenital lymphangioma circumscriptum was also reported². The tumour does not appear to have any particular predilection site and has been described in various corporal skin locations, subcutaneous tissue, but equally in deeper locations (e.g. brain, tongue, bone, spleen, testes)^{3–6}. We here present an exceptionally rare case of an adult occurrence of facial DT.

Case Report

A 63 year old woman presented with a 20 year history of slow-growing, small, right buccal dermatological lesion, which she thought was the common mole. On visual

inspection, the lesion had an unusual bluish black discoloration. According to the patient, she never suffered from any accompanying problems with this lesion (e.g. itching, bleeding) and only asked for the removal for cosmetic reasons. She was otherwise physically healthy, with no enlargement of local (or other) lymph nodes. There was no salient family or personal history. Following the discussion, in agreement with the patient, a wide local excision of 1.7x1x0.4 cm was performed and pathological examination done. Despite such a long history, the intradermal nodule was only 0.6 cm in maximum diameter (overall 0.5x0.6x0.1 cm in size). The analysis of formalin-fixed-paraffin-embedded (FFPE) and hematoxylin and eosin (H&E) stained biopsy revealed a tumour tissue beneath a normal looking epidermis. It was comprised of multiple delicate interconnecting vascular channels with papillary plugs which projected into the lumen lined by atypical plumped endothelial cells. Some of those plugs contained hyalinized core (Figures 1a–d). The distinct borders consisting of normal tissue with no further presence of tumour were present and the excision was deemed definitive. The histopathological diagnosis of DT was confirmed.

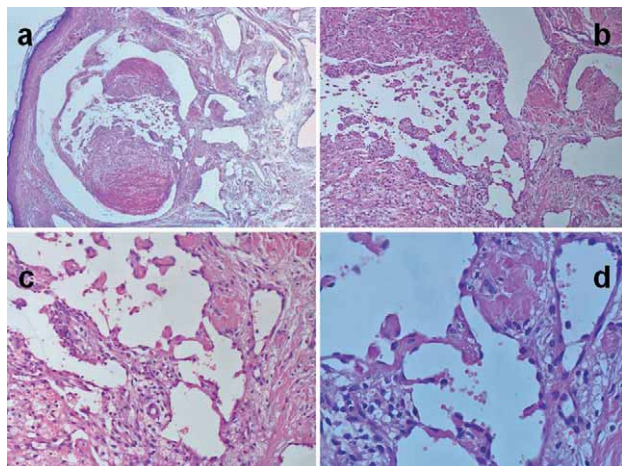


Fig. 1. a) The hematoxylin and eosin (H&E) stained biopsy revealed an intradermally located Dabska Tumour. The tumour tissues consisted of ectatic haemangioma-like/lymphatic-like capillaries (H&Ex40). b) Here, the formation of intraluminal papillary tufts almost obliterated cavernous space (H&Ex100). c) Delicate papillaries consisted of hyaline cores lined by prominent atypical nucleus and inconspicuous cytoplasm (H&Ex200). d) The typical hobnail or matchstick appearance of DT is shown (H&Ex400).

Discussion

DT is still considered controversial by some clinicians¹. However, due to a growing number of cases described in the literature^{1–6}, the accumulating knowledge of which we hope to add to with the case presented here, the consensus is being reached about the uniqueness and distinctiveness of this disorder. DT usually comes to medical attention when significantly bigger in size (approximately 2–3 cm in diameter) than was the case in our patient¹. It is reported to fluctuate in size and is commonly associated with degeneration of an underlying vascular tumour¹. In the adult case of facial DT that we report, despite a strikingly long history of growth (two decades), a careful and thorough examination and accompanying investigations revealed no metastases. The patient continues to be followed in the ENT outpatient clinic, with no reoccurrence of the lesion present in six months post-excision.

Acknowledgements

We are grateful to Dr Vladimir Bernić for the surgical expertise and support and to Dr Ivana Rosenzweig for the advice.

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DABSKA TUMOR U ODRASLE OSOBE: JEDAN NEOBIČAN MADEŽ

SAŽETAK

Godine 1969. Dabska i kolege prvi put su opisali ovaj rijetki maligni tumor, kasnije poznatiji kao maligni endovaskularni papilarni angioendoteliom dječje dobi. Danas se smatra kako tumor ima dobru prognozu, te da je široka ekscizija metoda izbora u liječenju ovisno o lokaciji. Ovdje prikazujemo rijedak i neobičan slučaj pacijentice u dobi 63 godine koja se javlja radi madežu slične, 20 godina prisutne, sporo rastuće dermatološke promijene desnog obraza. Dijagnoza Dabska tumora potvrđena je patohistološkom analizom hematoksilin & eozin (H&E) biopsije. Mikroskopska analiza otkrila je tumor građen od mnogobrojnih anastomozirajućih kavernozičnih kanala s papilarnim nakupinama među kojima pojedine resice, hijalinizirane vezivne strome, obložene atipičnim zdepastim endotelnim stanicama, prominiraju u lumen kapilara.