Original Article

Indian Journal of Pathology: Research and Practice

Histological Diagnosis of Angiofibroma in Tuberous Sclerosis

Jaisri Ananthapadmanaban

*Sr. Divisional Medical Officer, Dept. of Pathology, Southern Railway Headquarters Hospital, Perambur, Chennai-600 023, Tamil Nadu, India.

Abstract

Background: A ten year old male child presented with altered behaviour, low intelligence quotient and skin lesions. The child also had respiratory discomfort and palpitations. On investigation, a cardiac rhabdomyoma was located and excised. The skin lesions were ash leaf macules[1], shagreen patches and papulonodular lesions on the face and periungual region on the foot. This combination makes the diagnosis evident as tuberous sclerosis. **Methods:** Biopsy of the papulonodular lesions were done and histopathology revealed them as angiofibroma (adenoma sebaceum).[2] An angiofibroma by definition is supposed to have proliferation of blood vessels and fibrous tissue on microscopic examination. **Results:** This angiofibroma had in addition hyperplasia of sebaceous glands. A review of the literature revealed that sebaceous hyperplasia is only a secondary change in an angiofibroma. **Discussion:** This case is presented here to highlight the microscopic diagnosis of angiofibroma and to state that sebaceous gland proliferation though a secondary change, can also present in angiofibromas and pathologists need not look for another diagnosis.

Keywords: Angiofibroma; adenoma sebaceum; sebaceous hyperplasia.

Introduction

A ten year old male child presented with low intelligence, altered behaviour and skin lesions. He also had palpitations and respiratory discomfort. He was investigated and echocardiography revealed a tumour in the left atrium. It turned out to be a cardiac rhabdomyoma[3] and was excised. The skin lesions were ash leaf macules, shagreen patches and papulonodular lesions on the face and periungual region. Biopsy of the papulonodular lesion on the periungual region of the great toe was diagnosed as angiofibroma (adenoma sebaceum).[4] The child was diagnosed to have tuberous sclerosis with all the clinical features put together.

Tuberous sclerosis is an autosomal dominant neurocutaneous syndrome in which mental retardation and epilepsy with angiofibromas are present. Multiple hamartomas involving other organs can also be present. The cutaneous lesions[5] constitute angiofibromas, shagreen patches and ash leaf macules. The angiofibromas are present on the nasolabial folds, cheeks, chin, face, scalp, subungual and periungual regions. They present as small red smooth papules or soft brown nodules. Thus

Corresponding Author: Jaisri Ananthapadmanaban, T 87/A, 29th Cross Street, Besant Nagar, Chennai 600090, Tamil Nadu, India.

E-mail: jaisri12@yahoo.com

© Red Flower Publication Pvt. Ltd.

the clinical diagnosis of tuberous sclerosis[6] was made.

Histopathological findings[7]

The predominant findings are proliferation of fibroblasts in the dermis which are stellate in shape and capillaries with some showing dilatation. The epidermis shows flattening of rete ridges and mild hyperkeratosis. A sparse inflammatory infiltrate is sometimes observed. Contrary to the findings of atrophic sebaceous glands, this lesion showed hyperplasia of the sebaceous glands in addition to the fibrous and capillary proliferation. A search of the literature revealed only one article by Sanchez, Wick and Perry[2] which says that the proliferation of sebaceous glands is only a secondary event in an angiofibroma. This must have been the reason for the misnomer 'Adenoma sebaceum' of Pringle in earlier days. Elastic tissue was absent from the stromal fibrous tissue.

Materials and Methods

The biopsied skin covered soft tissue was 0.5x0.5x1 cms in dimensions. It was all embedded. Sections from the paraffin wax blocks were stained with Hematoxylin and eosin stain and coverslipped. The histopathological features were studied and observed. Further sections from the same paraffin wax blocks were stained with Verhoeff Van gieson's stain to demonstrate stromal fibrosis and elastic tissue and the features were observed.

Observation

The Hematoxylin and eosin stained sections revealed the proliferation of fibroblasts and capillaries in the dermis. There was hyperplasia of the sebaceous glands in addition. The Verhoeff Van gieson's stain confirmed the proliferation of fibrous tissue and collagen fibres and the absence of elastic tissue in the dermis.

Discussion and Conclusion

Angiofibromas of the face is one of the cutaneous features[8] of tuberous sclerosis; it can occur in other conditions like neurofibromatosis 2 (NF-2), multiple endocrine neoplasia (MEN) type I and perifollicular angiofibromas occur in Hornstein-Knickenberg syndrome. The differential diagnosis of angiofibroma is solitary angiofibroma of the face or fibrous papule of the face[9] or nose from which it is indistinguishable except from the fact that the vessels are more ectatic[10] and less likely to show concentric fibrosis than in adenoma sebaceum.

The differential diagnosis of angiofibroma [11] with sebaceous hyperplasia as a secondary feature would be folliculosebaceous cystic hamartoma[12], sebaceous trichofolliculoma and nevus sebaceous. In folliculosebaceous cystic hamartoma, the predominant component is the sebaceous hyperplasia and cystic infundibular structures with radiating sebaceous lobules are present. The fibrous and vascular components are admixed with adipose and neural tissue in varying proportions. Angiofibromas of tuberous sclerosis which exhibit sebaceous hyperplasia, has it only as a minor component and the cystic infundibular structures are lacking. Sebaceous trichofolliculoma possesses sebaceous hyperplasia in addition to proliferation of follicles which are absent in angiofibromas. Nevus sebaceous of Jadassohn shows proliferation of other adnexal structures like apocrine and eccrine glands, a slight increase in vascularity and epidermal changes in addition to sebaceous gland hyperplasia. The epidermal changes include acanthosis and papillomatosis. All these lesions occur commonly on the face. Other histological similarities would be senile sebaceous hyperplasia, the sebaceous glands of rhinophyma and shallow shave biopsies which would show prominent sebaceous glands. These lesions may not be accompanied by the proliferation of fibrous stroma and capillaries

as in angiofibroma and hence can be differentiated. Moreover, the clinical history and other coexisting lesions would go hand in glove in making the correct diagnosis.

References

1. Fitzpatrick TB,Szabo G, Hori Y *et al*. White leaf shaped macules. *Arch Dermatol*. 1968; 98: 1.
2. Sanchez NP, Wick MR, Perry HO. Adenoma sebaceum of Pringle: A clinicopathologic review, with a discussion of related pathological entities. *J Cutan Pathl.* 1981; 8: 395.
3. Moreles JB, Congenital rhabdomyoma, tuberous sclerosis and splenic histiocytosis. *Arch Pathol.* 1961; 71: 485.
4. Park YK, Hann SK. Cluster growths in adenoma sebaceum associated with tuberous sclerosis. *J Am Acad Dermatol.* 1989; 20: 918.
5. Reed RJ. Cutaneous manifestations of neural crest disorders (neurocristopathies). *Int J Dermatol.* 1977; 16: 807-826.
6. Schnur RE. Tuberous sclerosis. The persistent challenge of clinical diagnosis. *Arch Dermatol.* 1995; 131: 1460-1462.
7. Nickel WR, Reed WB. Tuberous sclerosis; special reference to the microscopic alterations in the cutaneous hamartomas. *Arch Dermatol.* 1962; 85: 395-403.
8. Webb DW, Clarke A, Fryer A, Osborne JP. The cutaneous features of tuberous sclerosis. *Br J Dermatol*. 1996; 135: 1-5.
9. Meigel WN, Ackerman AB. Fibrous papule of the face. *Am J Dermatopathol.* 1979; 108: 673-675.
10. Reed RJ, Hairston MA, Palomeque FE.The histologic identity of adenoma sebaceum and solitary melanocytic angiofibroma. *Dermatol Int.* 1966; 5: 3-11.
11. Bhavan J, Edelstein L. Angiofibromas in tuberous sclerosis; a light and electron microscopic study. *J Cutan Pathol.* 1977; 4: 300-307.
12. David Weedon. Tumours of cutaneous appendages. Skin Pathology, 2nd Ed. 2002: 871-873.

Red Flower Publication Pvt. Ltd,

CAPTURE YOUR MARKET

For advertising in this journal

Please contact:

International print and online display advertising sales

E-mail: redflowerppl@vsnl.net / tel: +91 11 22754205, 45796900

Recruitment and Classified Advertising

E-mail: redflowerppl@vsnl.net / tel: +91 11 22754205, 45796900

Disclaimer The opinion in this publication is those of the authors and is not necessarily those of the **Indian Journal of Pathology: Research and Practice** the Editor-in-Chief and Editorial Board. Appearance of an advertisement does not indicate **IJPRP** approval of the product or service.