CASE REPORT

NAEVUS SEBACEOUS OF JADASSOHN'S OF EYELID

Alyscia M. Cheema, Nausheen Hayat
Department of Opthalmology, Jinnah Post Graduate Medical Centre, Karachi-Pakistan

Naevus Sebaceous of Jadassohn is a rarely seen hamartomatous lesion and is a fourth type of sebaceous gland tumour, in which sebaceous glands show nevoid character of growth composed partly or completely of sebaceous glands. A detailed research revealed that very little data is available; no case of this disorder has been reported as an isolated eye lid lesion. Majority of the published cases are associated with systemic involvement. We present here a case report of this rare disorder. A middle age female patient presented with a mass of left lower lid since childhood without any other ocular and systemic abnormality. Excision biopsy along with rotation flap surgery was done. Histopathological examination of the specimen revealed the diagnosis. To our best of knowledge this is a unique presentation of naevus sebaceous of Jadassohn on eye lid which has not been documented till yet anywhere.

Keywords: Eyelid, naevus sebaceous, Jadassohn, Hamartoma

J Ayub Med Coll Abbottabad 2015;27(4):927–9

INTRODUCTION

In 1895 German dermatologist Joseph Jadassohn first highlighted a syndrome by name of Naevus Sebaceous to narrate new growths involving partial or complete sebaceous gland with characteristic nevoid appearance.[1] Later on Robinson proposed a term of Naevus Sebaceous Jadassohni for it.[2]

Most of the cases occur sporadically with occasional reports of familiar cases. Sebaceous nevi can be found in 0.3% of infants. The sex incidence is more frequent in females as compared to males with ratio of 2:1.[3] Common sites of occurrence are the scalp, retroauricular areas, temples, forehead and central part of the face. We have not found any reports of sebaceous nevi on the lid in the literature.

CASE REPORT

A fifty year old female patient presented to the eye O.P.D. of Jinnah Post-Graduate Medical Centre (JPMC) with a growth on the left lower lid. The growth was there since childhood with gradual progressive increase in size. On examination, there was a pinkish grey warty growth, 20×25 cm, involving 80% of the lower lid, non-tender, adherent to the tarsal plate, which was firm in consistency. No lymph adenopathy was found .3 years ago cautery was tried on her lesion in village. There were multiple black pigmented lesions on forehead and both medial canthi. (Figure-1).

On examination patient had visual acuity of 6/6 and N6 with +1.50 DS in both eyes. Anterior and posterior segment normal. IOP was 10 mm Hg. There was no other systemic abnormality present. No significant family history present.

A diagnostic biopsy was done. The lesion revealed hyperkeratosis, acanthosis and areas of hypergranulosis and papillomatosis in the epidermis. The dermis has abnormally developed her follicles with prominent groups of numerous sebaceous glands, along with scattered apocrine glands. There was no evidence of any associated neoplasm (Figure-2 & Figure-3). It was decided to re-sect the lower lid, followed by lid reconstruction (Figure-4)

The patent was infiltrated with local anaesthesia, after making the preoperative skin markings, 80% of the lower lid had had to be excised. The resultant defect was reconstructed using a temple and cheek rotation flap (Mustarde). The patient had excellent functional and cosmetic results (Figure-4).

DISCUSSION

There is a wide variety of structural diversity in the eyelid. The eyelid contains apocrine and eccrine sweat glands, hair follicles and sebaceous glands along with tarsal plates, muscles, nerves and blood vessels. All of them contribute to the integrity of the eyelid. Anyone of these structures may undergo tumefaction.[4] Benign tumours are one of the most frequently occurring lesions on the eyelids.[5,6]

It is important while evaluating the eyelid lesions that many malignant tumours can mimic the appearance of a benign lesion. Early diagnosis requires an accurately taken history, a high index of suspicion and most important a diagnostic biopsy should be performed.

Naevus sebaceous of Jadassohn is harmatomatous organoid malformation or sebaceous glands predominantly, along with apocrine glands, hair follicles and the epithelial layer.[7]

.

It usually present at birth as a yellowish orange area.At time of puberty the lesion becomes yellow to dark brown,verrucoid and is micrnodular.[8]

The characteristics histology[9] is usually seen at puberty consisting of mature sebaceous glands and occasional papillomatous hyperplasia of the overlying

epidermis. The hair follicles are usually inconspicuous. Ectopic apocrine glands are commonly seen deep in the dermis beneath the sebaceous glands. Life history of naevus sebaceous through an early phase (infancy & childhood).[8,10]

With a typical lesion as a flesh coloured plaque on the scalp or the face. The overlying epidermis may be normal or show mild papilomatous hyperplasia in the second phase the alopecic area becomes a yellow waxy papilllomatous plaque as a result of the development of sebaceous and apocrine glands. The skin surface is formed by soft, yellow fibroepithelial papillomas. In the third phase the development of secondary hyperplasia or neoplasia of the surface epidermis pilosebaceous structures and apocrine glands marks this phase.

Rapid circumscribed enlargement, ulceration or development of an exophytic nodule should arouse suspicion of malignant transformation which is a well-recognized complication.Tumor, in order of frequency are Basal Cell Carcinoma, Adenexal and pilar tumors 40% each and Apocrine neoplasms 20%

Systemic Malformations Associated with Naevus Sebaceous are Epilepsy,Mental Retardation and Skeletal Deformities.The associated ophthalmic findings are Corneoscleral Limbal Choristomas,Intrascleral Cartilage and Bone,Retinal Detachment, Optic Nerve Colobomas and Microphthalmos[11,12]

Figure-1: Preoperative photograph showing lesion on left lower lid

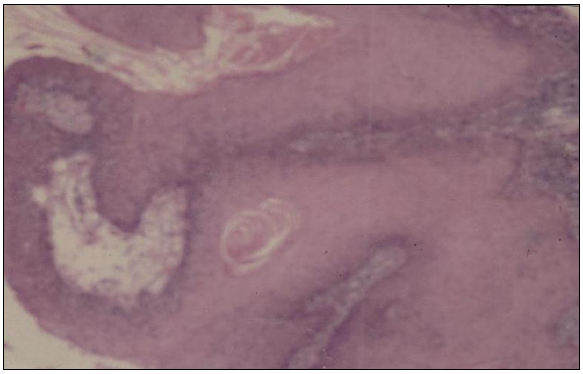

Figure-2: 150X, microscopic photograph of epidermis showing hyperkeratosis, acanthosis and papillomatosis

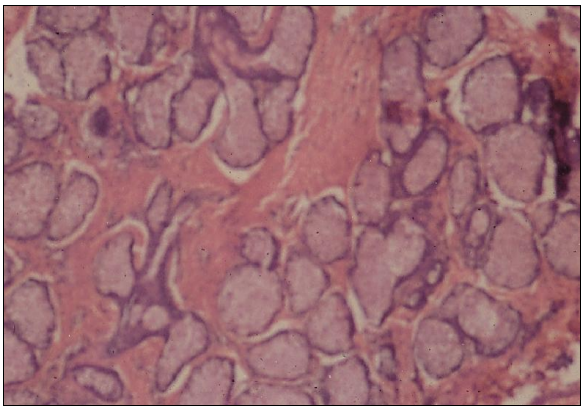

Figure-3: 400X, microscopic photoghraph of dermis showing lobules of sebaceous glands, hair follicles and ectopic apocrine glands

Figure-4: Post-operative

ACKNOWLEDGMENT

Both of the authors worked together in diagnosis, management and writing this case report.

We would like to express our regards and thanks to Dr. Saleem Sadiq, histopathologist by profession, who helped us in making this diagnosis. Both authors have no potential conflicts of interest including relevant financial interests, activities, relationships, and affiliations regarding this study, as well as declarations of no conflicts of interest, must be included in the Acknowledgment section of the manuscript.

REFERENCES

1. Robinson SS. Naevus sebaceous (Jadassohn) report of four cases. Arch Derm Syphilol. 1932;26(4):663–70.
2. Zaremba J. Jadassohn's naevus phakomatosis: 2. A study based on a review of thirty-seven cases. J Ment Defic Res 1978;22(2):103–23.
3. Serpas de López RM, Hernández-Pérez E. Jadassohn's sebaceous nevus. J Dermatol Surg Oncol 1985;11(1):68–72.
4. Lucas DR. Greers Ocular Pathology, 4th Edition. Blackwell Scientific Publications, 1989;6:81–97.
5. Aurora AL, Blodi FC. Lesions of the eyelids: A clinicopathological study. Surv Ophthalmol 1970;15(2): 94–104.

6. Saeed M. Results & discussion in tumors of the eyelid & Surgical Management of basal cell carcinoma. Dissertation. CPSP, Pakistan-1990; 83
7. Atherton DJ. Naevi and other developmental defects. In: Champion RH, Burton JL, Ebling FJ. editors. Rook/Wilkinson/Ebling Textbook of Dermatology. 5th Ed. Oxford: Blackwell Scientific Publications; 1992. p.445–526.
8. Labbé D, Badie Modiri B, Petit F. Sebaceus nevus of Jadassohn. Apropos of 62 surgically treated cases and review of the literature. Rev Stomatol Chir Maxillofac 1999;100(4):175–9.
9. Kao GF. Benign tumors of the epidermis in Pathology of the skin. Farmer ER, Hood AF, eds. Norwalk, CT, Appleton & Lange 1990; p.534–6.
10. Puri N. A clinical and histopathological study of nevus sebaceous. J Pak Assoc Dermatol 2014;24(1):31–3.
11. Albert D, Miller J. Albert & Jakobiec's Principles & Practice of Ophthalmology. vol. 4. Phila WB Saunders Co 2000;3238–9.
12. Patrizi A, Pizzino D, Tullini A, Gobbi G, Santucci M. Sebaceous linear nevus syndrome with hemimegaloencephaly. Report of a case. G Ital Dermatol Venerol 1989;124(1-2):55–8.

Address for Correspondence:

Dr Nausheen Hayat, Eye Department, Jinnah Post Graduate Medical Centre, Karachi-Pakistan
Cell: +92 321 210 1888
Email: nausheen_hayat@hotmail.com