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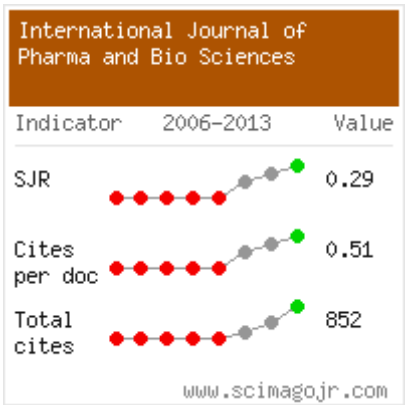

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Case Report **Microbiology**

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A CASE OF LOBULATED CAPILLARY HEMANGIOMA OF THE NOSE WITH LINEAR AND WHORLED NEVOID HYPERMELANOSIS

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ABSTRACT

Nasal lobular capillary hemangioma (LCH) is a rare tumor of the nose and paranasal sinuses. The exact etiopathogenesis is still unknown. Linear and whorled nevoid hypermelanosis (LWNH) is also a rare skin condition. Both are associated with chromosomal and systemic anomalies. A case of a 24 year old female patient who presented with epistaxis and nasal obstruction is reported. She was found to have associated LWNH. The various etiological factors and management are discussed. This is a unique case and first of its kind reported in the literature.

KEY WORDS: **Lobular capillary hemangioma, Linear hypermelanosis, chromosomal anomalies.**

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INTRODUCTION

Hemangioma comprises of a heterogeneous group of benign proliferation of blood vessels developing during childhood. There are two forms: capillary and cavernous. The capillary form consists of numerous small capillaries. Cavernous hemangioma is of large dilated sinuses filled with blood[1]. LCH is a specific lesion among the vascular lesions of the head and neck. Nasal lobular capillary hemangioma is a rare benign tumour of the nose and paranasal sinuses[2]._LWNH is a rare sporadic pigmentary skin condition characterized by swirls and streaks of macular hyperpigmentation along the Blaschko line appearing within the first two years of life[3]. It is occasionally associated with chromosomal abnormalities and anomalies of other organs. Both these conditions appear early in life, and may be congenital. We are reporting such a case who presented at our ENT casualty with epistaxis and to the best of our knowledge, this is the first case to be reported in the literature.

CASE REPORT

A 24 year old female presented at the casualty with moderate epistaxis from the right nasal cavity. She gave a history of progressive nasal obstruction for the past 6 years and recurrent nasal bleed during the last 5 months. General examination revealed linear, whorled hyperpigmented macules on both upper and lower limbs, and trunk which were more on the left side, and were present from the first few weeks of birth. It was diagnosed clinically as LWNH by the dermatologist (Fig 1).

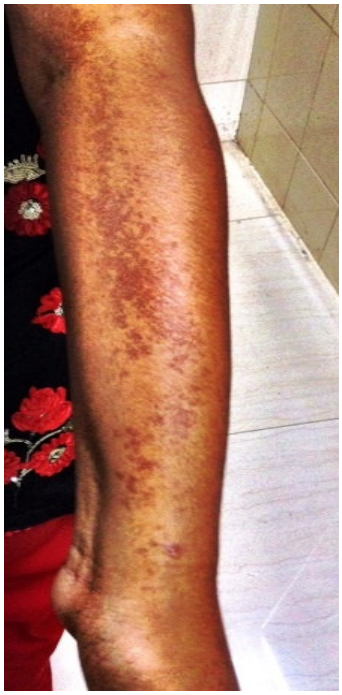

Figure 1
Linear and whorled hyperpigmentation on the left upper limb sparing the palm

On diagnostic nasal endoscopic examination, a pale pink polypoidal mass was found filling the posterior half of the right nasal cavity. The mass was smooth, lobulated, filling the right choana into the nasopharynx and jutting into the left choana. CT scan of the nose and paranasal sinuses showed a homogenous soft tissue density mass in the right nasal cavity

abutting the inferior turbinate laterally and middle turbinate superiorly. On MRI, the lesion was hyperintense in T2 and hypointense in T1 (Fig 2). Her coagulation profile, LFT, TSH and RFT values were normal. Systemic examination including the cardiac, skeletal and ocular systems was normal.

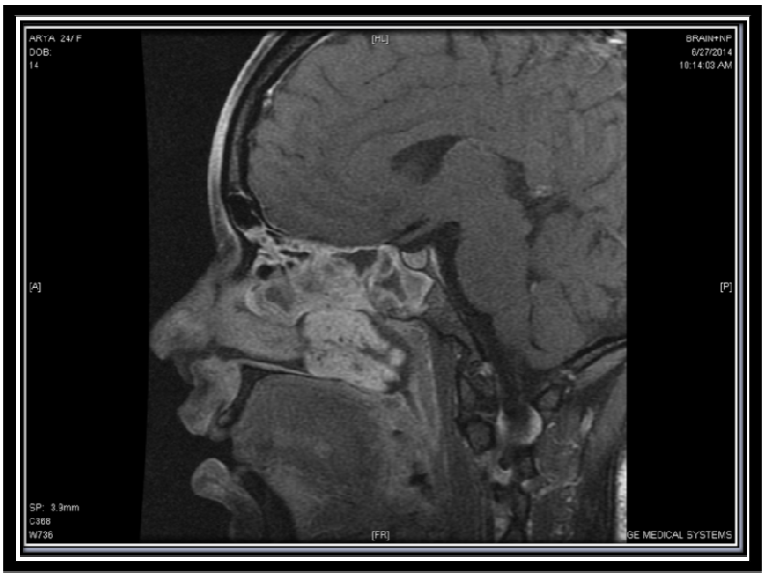

Figure 2
MRI scan-Well enhancing lesion in the posterior part of the nasal cavity extending into the choana

Under general anesthesia, the nasal mass was removed endoscopically. It was found to be attached to the right inferior turbinate with the posterior part eroded and with indentation on the middle turbinate. The mass measuring 4×2.5×1.5cm was firm in consistency (Fig 3). There was profuse bleeding which was controlled by a tight anterior nasal packing. Post operative period was uneventful.

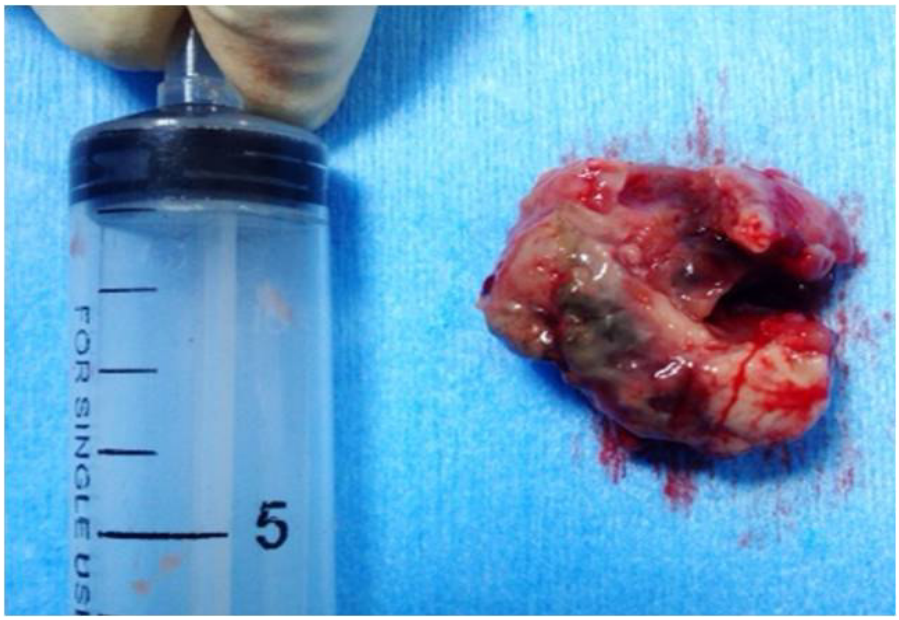

Figure 3
Excised mass from the nose

Microscopically, the lesion was covered by pseudo-stratified epithelium. The underlying stroma showed dense fibrosis and lobules of small capillaries with chronic inflammatory infiltrate of lymphocytes, eosinophils, plasma cells, and neutrophils. On immunohistochemistry, CD 34+ positive endothelial cells were present (Fig 4). These findings led to a diagnosis of lobular capillary hemangioma.

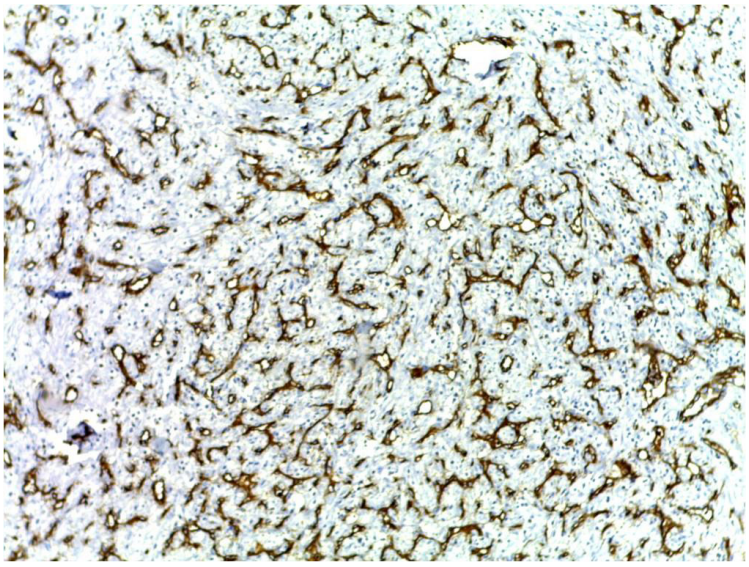

Figure 4
CD34+ endothelial cells by immunohistochemistry

LWNH was confirmed by skin biopsy of the pigmented lesion. On follow up, no recurrence has been noted during the last 6 months.

DISCUSSION

First described in 1897 by two French surgeons, Poncet and Dor, who named this lesion botryomycosis hominis [2], the term pyogenic granuloma is a misnomer as it is not a true granuloma nor a pus producing lesion. In 1980, Mills et al. termed pyogenic granuloma as lobular capillary hemangioma because of its characteristic microscopic features. First case of pyogenic granuloma of the nasal cavity was described in 1940 by Frank I, and Blahd M[2]. LCH has also been referred to by other names such as, granuloma gravidarum, hemangiomatous granuloma, granuloma telangiectacticum and also Crocker and Hartzell's disease [4]. Lobular capillary hemangioma occurs from 10 months of age to the 7th decade of life. Most common site is the head and neck region. Females are more affected. Commonly they are seen on the lips, oral cavity and tongue [5]. Rarely the eye, trachea and nose are involved[6]. Nasal lobular capillary hemangiomas generally arise from the nasal septum, turbinates, on the roof of the nasal cavity or in the maxillary sinus[7]. Clonality has been demonstrated cytogenetically in a case from the nasal cavity, showing deletion within the long arm of chromosome 12[8]. Capillary hemangiomas have been associated with ocular and arterial anomalies and posterior fossa malformations[9]. Histologically, the LCH lesion consists of two areas, lobular and superficial ulcerative. The lobular area is characterized by capillary proliferation with a microscopically distinct lobular architecture. The superficial portion of the lesion may undergo nonspecific changes, including stromal edema, capillary dilation, inflammation, and a granulation tissue reaction[7]. Linear and whorled nevoid hypermelanosis (LWNH) coined by Kalter in 1988, persist throughout life and is thought to reflect mosaicism or chimerism. Chromosomal anomalies of LWNH include mosaic trisomy 7, 14, 18 and X-chrosomal mosaicism[3]. It may be associated with growth retardation, body asymmetry, ocular and cardiac, central nervous and musculoskeletal anomalies. Histology shows epidermal melanosis. Association with capillary hemangioma has not been reported. Contrast enhanced CT features of a LCHconsist of an intensely enhancing mass and an iso or hypoattenuating cap of variable thickness around it, with or without associated bony destruction. Studies have shown bony erosion to be associated with LCH[7]. A differential diagnosis of a hypervascular mass of the nasal cavity in patients with nasal obstruction and or epistaxis might include juvenile angiofibroma, angiomatous polyp, hemangioma, hemangiopericytoma, paraganglioma, angiosarcoma, and hypervascular metastases, particularly from kidney, thyroid, lung, or breast. None of these lesions have been reported to have an iso- or hypoattenuating cap of variable thickness around the intensely enhancing lobular mass in the literature [7.] Excision of the entire lesion endoscopically is the treatment of choice. Promising results have been reported for various new treatments, such as laser therapy and sclerosing agents[3.] Regarding recurrence rate, Mills et al observed a low recurrence rate of 11.1%, while Steven C. Smith, Rajiv M. Patel et al reported a recurrence rate of 42%[9]**.** Recurrence is usually due to incomplete excision[8]. The narrow space with a rich vascularity of the nasal cavity are factors to be considered leading to recurrence[9]. Nowadays, with nasal endoscopy, better visualization may help to bring down the recurrence rate**.** No malignant transformations have been reported[10]. Chemical peeling has been tried with trichloroacetic acid for LWNH, with no benefit[11]. In the present case, the patient had developed linear streaks of macules in the first few weeks of life and was diagnosed with LCH at the age of twenty one. She did not have any associated systemic anomalies, though cytogenetic studies were not done. LCH with LWNH has not been reported in the literature before.

CONCLUSION

This case report draws attention to the uncommon location of the LCH and the possible association with LWNH. Both these

conditions have chromosomal cytogenetic anomalies. Systemic anomalies have also been associated with them. Further immunocytogenetic studies should be carried out to find the correlation between these two uncommon conditions.

CONFLICT OF INTEREST

There is no conflict of interest.

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