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Dermatology

ANGIOBLASTOMA OF NAKAGAWA(TUFTED ANGIOMA) IN EAR AURICLE: A RARE CASE REPORT

Dr. Amar Singh*	MBBS, MD DVL, Senior Resident, Department of Dermatology, Venereology & Leprosy, Autonomous State Medical College, Shahjahanpur, UP *Corresponding Author
Dr. Mamta Verma	MBBS, DNB ENT, Senior Resident, Department of Otorhinolaryngology, Autonomous State Medical College, Shahjahanpur, UP
Dr. Astha Pant	Consultant Dermatologist, Government Combined Hospital, Dehradun, Uttarakhand

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INTRODUCTION:

Angioblastoma of Nakagawa or acquired tufted angioma is a benign, slow growing vascular proliferation commonly localized in skin and subcutaneous tissue with typical histological features. They mostly develop in infancy or early childhood, with a protracted course, but it can also occur in adulthood, as the case reported in this article.

CASE REPORT:

23 years old male patient presented to Dermatology OPD with complain of gradual increase in size of left ear auricle since 6 years. He stated that it started as single red spots over pinna which progressed to whole ear. There was mild itching, pain on touch and occasional localized sweating. He had no previous history of trauma, treatment or any other co morbidities.

CUTANEOUS EXAMINATION:

Increase in volume of left ear auricle, well defined erythematous, indurated, telengiectatic plaque with almost involving whole of ear auricle with irregular contours. There were 3 erythematous papule over anterior border of helix.The local temperature was raised, slight tenderness was present. Pulsation due to underlying blood vessels was noted (Figure 1and 2).

HISTOPATHOLOGICAL EXAMINATION:

Biopsy shows numerous thin walled dilated capillaries in the upper and mid dermis. Most of these showed complete lining of endothelial cells and contained RBCs and plasma. A thick walled venule was also seen in mid dermis (Figure 2and 3).

DISCUSSION :

Angioblastoma of Nakagawa or acquired tufted angioma is a benign vascular tumor. Most cases occur in early childhood, although adult cases has also been reported. Around 25% tufted angiomas are congenital and 50% occurs in first year of life. There is no sex predilection. Origin is lymphatic based on immuno staining pattern, otherwise etiopathogenesis is unknown.

It shows many clinical patterns on neck, trunk, abdomen and limbs. They may presents as subtle stain like area which progress to plaque like, infiltrated, red or blue-purple lesion and as exophytic rubbery to firm, vialeous cutaneous nodule. There may be mild itching, tenderness and hyperhidrosis in some cases. They are usually solitary, but multifocal cases have also been reported[1].

Tufted angiomas can regress spontaneously within few years and can remain unchanged. Tufted angiomas that are present at birth or in first year of life have a greater tendency to spontaneously regress than those that appear later in life[2].

Kasabach- Merritt phenomenon can develop in tufted angiomas.

Histopathologicaly there is tuft of tightly packed capillaries dispersed in dermis in a typical cannonball distribution with lymphatic spaces in tumor stroma and cresentic spaces around vascular tufts[3].

No definite treatment guideline for management till now. Recommended therapies are compression, excisions, topical or systemic corticosteroids, cryotherapy, embolization ,various lasers, propranolol and chemotherapy[4][5][6][7].

Cutaneous lesion in ear auricle are present in different pathology, early recognization of lesion is required for further management. Differential diagnosis include infantile hemangioma, venous malformation, congenital hemangioma, Kaposi's sarcoma, lepromatous leprosy, sarcoidosis, polymorphic light eruption, leishmaniasis, lacaziosis, chronic perichondritis, cellulitis, granulomatosis with polyangitis, chondrodermatitis nodularis helicis. Biopsy is always required to confirm the diagnosis.

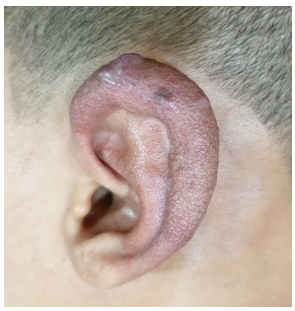

FIGURE 1: Increase in volume of right ear auricle, well defined erythematous, indurated, telengiectatic plaque with almost involving whole of ear auricle with irregular contours. There were 3 erythematous papule over anterior border of helix.

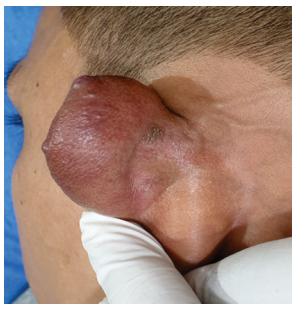

FIGURE 2: Erythematous plaque over helix and extending to whole pinna.

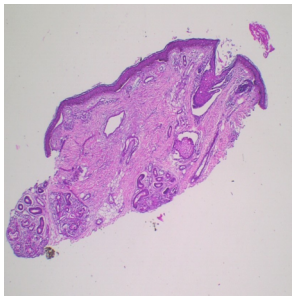

Figure 3: Biopsy Shows Numerous Thin Walled Dilated Capillaries In The Upper And Mid Dermis

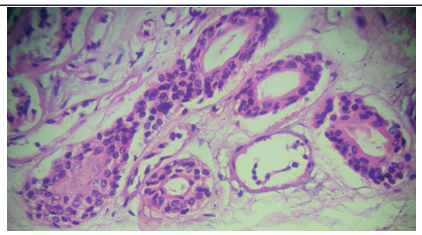

Figure 4 : Higher Magnification 10×40 Shows Prolifertating Capillaries

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