Neurovascular Hamartoma of Face: An Unusual Clinical Presentation

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ABSTRACT

Introduction: Neurovascular hamartomas (NVH) are relatively uncommon and have been very rarely reported in head and neck region. The rarity could be attributed to it being unrecognized and/or unreported. They constitute unique group of lesions with specific histopathological features.

Case report: Authors report a case of 23-year-old male patient with NVH of face with an unusual clinical presentation.

Conclusion: Neurovascular hamartoma is a histopathological diagnosis made for small to medium-sized vessels and closely packed groups of well-formed nerve bundles within a loose connective tissue matrix and the inflammatory component is minimal or absent.

Clinical significance: Neurovascular hamartomas have specific features, and, therefore, they should not be considered as any reactive lesion or any hamartomatous lesions. Neurovascular hamartomas in fact represents a separate and unique entity.

Keywords: Face, Hamartoma, Neurovascular.


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INTRODUCTION

Albrecht in 1904, first introduced the term “hamartoma” derived from a Greek word hamartia meaning “character flaw”. He described it as tumor-like but nonneoplastic malformations or errors of tissue development, which results in abnormal admixture of tissue indigenous to the site. Hamartomas can arise from any of the three germ layers with any type of tissue as a predominant tissue one. It can occur at a number of sites as it is considered as developmental error. Histologically, there is difference between hamartomas, choristoma, teratomas, and dermoids. A hamartoma consists of abnormal proliferation of tissues, which are endogenous to that site. This is in contrast to choristoma, which consists of abnormal proliferation of tissue exogenous to the occurring site. Whereas teratomas consist of pluripotent tissue, i.e., tissue derived from all three germ cell layers, and this tissue is often foreign to the site of occurrence. On the contrary, dermoids are usually cystic. They contain either ectodermal or mesodermal elements, primarily the former. Among these, the highest malignant transformation rate is of teratomas. In choristoma, it is <1%. Hamartomas have little or no malignant transformation rate. According to the predominance of the tissue, hamartomas can be classified as either vascular angiomatous, lipomatous, chondroid, osseous or neurogenic. Hamartomas are relatively common in the skin, lung, liver, chest wall, kidney, and gastrointestinal tract, but uncommon in the region of the head and neck as they present diverse clinical and microscopic features. Neurovascular hamartomas (NVH) have been very rarely reported in head and neck region owing to it being unrecognized, and, therefore, frequently unreported. Here, we report a case of NVH of face and discuss its clinical presentation and histopathological differential diagnosis.

CASE REPORT

A 23-year-old male patient visited the Department of Oral and Maxillofacial Pathology and Microbiology with the chief complaint of facial disfigurement on left side of face. History revealed that the patient noticed change in the facial skin 7 years ago. It gradually increased to an irregular mass. Extraoral examination revealed a massive, baggy, and pendulous mass of skin on left side of face (Fig. 1). It extends from lower eyelid to 0.5 cm below the inferior border of mandible. Anteroposteriorly, it extends from left ala of nose to external auditory meatus. The growth had displaced the outer canthus of left eye upward. The skin over the growth is thrown into...
irregular folds. At places, it revealed orange peel appearance. The color of the skin was also darker as compared to normal adjacent skin. Tenderness was present. It was soft to firm in consistency. Incisional biopsy was done and sent for histopathological examination.

Histopathological examination revealed epidermis along with underlying skin appendages. The lesional tissue was not clearly demarcated from the adjacent normal tissue. It was composed of diffuse proliferation of variously sized matured thick and thin-walled endothelial lined blood vessels interspersed with the loose fibroadipose tissue (Figs 2 and 3). These blood vessels were grouped in close proximity to well-formed nerve bundles (Fig. 4). Most of the blood vessels were filled with red blood cells. The relative density of vascular component seemed more dominant. Mild chronic inflammation was evident at places. The presence of nerve tissue in close proximity to blood vessels along with fibroadipose tissue in loose connective tissue stroma favored the final diagnosis of NVH. The growth was then excised, which showed similar findings. Facial contouring was done. Healing and postoperative course was uneventful. No recurrence was evident after 6 months of follow-up.

DISCUSSION
Currently, in literature, no specific and clear diagnostic criteria have been laid down for NVH. The term hamartoma is defined as nonneoplastic malformation characterized by proliferation of mature cells and tissues indigenous to the affected part. However, there is an excess of one or more tissue types in a disorganized manner. According to some sources, these hamartomas often tend to be present at birth or in young age but certain cases have also been reported in later age of life.
Therefore, it can be clearly stated that the diagnosis of hamartomas cannot be made just on age factor.

There are various sites of occurrence of hamartomas in head and neck region, which include the nasopharynx, larynx, middle ear, Eustachian tube, and recently the oral cavity, which included tongue, buccal mucosa, and lower lip. However, NVH of skin of face is rarely documented. Only two such cases have been previously published by Perez-Atayde et al who reported congenital NVH of the skin as a possible marker for malignant rhabdoid tumor.

The lesion here is unusual in the sense that it was histopathologically composed of numerous thick and thin-walled mature blood vessels of varying size, along with several well-formed nerve tissue embedded in fibrofatty stroma. These features are similar to NVH as confirmed by morphometric analysis of Allon et al. They stated that the two features, which can be used as diagnostic criteria of NVH are presence of several recognizable nerve bundles and nerve bundles grouped in close proximity to blood vessels.

Clinically, this case was difficult to distinguish from neurofibromatosis as it also sometimes manifests with massive overhanging masses. Neurofibromatosis may be present at birth but it often begins to appear during puberty and continue to develop slowly. However, a highly characteristic feature is the presence of cafe au lait pigmentation on skin. These were absent in present case. Moreover, histopathologically, it is composed of interlacing bundles of spindle shaped cells with wavy nuclei and numerous mast cells. This case showed numerous blood vessels along with nerve tissue.

Another clinical differential diagnosis includes Proteus syndrome (PS). Proteus syndrome is a complex hamartomatous disorder defined as local overgrowth (macrodactyly or hemihypertrophy), subcutaneous tumors and various bone, cutaneous and/or vascular anomalies. It is most commonly presented in children. In this case, apart from hemihypertrophy of face, no other features were present, therefore, the diagnosis of PS was excluded.

The histopathological differential diagnosis for NVH includes hemangioma which is often difficult to distinguish from it. Vascular hamartoma or NVH are typically differentiated from hemangiomas by a careful history and physical examination. Hamartomas are commonly asymptomatic and even if present at birth, they may not be apparent until adolescence or adulthood as in present case. However, capillary hemangiomas typically become apparent in the 1st week of life and rapidly proliferate in infancy only to involute in early childhood. Moreover, histopathologically, hemangiomas either lack or show minimal neural component.

The distinction between NVH and traumatic neuroma also presents a diagnostic dilemma. Traumatic neuroma, especially, can be difficult as sometimes, it may also show vascular component. The distinction is primarily based on location of the vascular proliferation. In case of NVH, the vascular component is intermingled with the neural component, whereas in case of neuroma, the vascular and the neural components are separate and the neural component predominants. The presence of reactive features, such as inflammation and fibrosis are more suggestive of a traumatic etiology. In this case, inflammation was minimal. Traumatic neuroma can be differentiated from NVH by lack of sensory or pain disturbances in the latter. However, in this case, tenderness may be elicited because of compression of nerves by the lesional tissue.

Despite their association with malignant tumors, such as adenocarcinoma of colon and neurogenic sarcomas; hamartomas are self-limiting benign lesions. However, two cases of congenital NVH of skin have been reported to precede malignant rhabdoid tumor. The neural component consists of bland spindle cells. Whereas, in this case, well-formed nerve bundles were observed.

No single treatment modality is favored in NVH. Surgery, laser therapy, and sclerotherapy are all options for treating the vascular malformations. Surgical excision is appropriate for localized lesions with fibrofatty remnants in cases of involuted hemangiomas. Elective subtotal excision of massive protuberant proliferating hemangiomas can be employed to maintain esthetic facial boundaries. The NVH should be treated similar to hemangiomas and vascular malformations. The combination of treatment options should be utilized. Sclerotherapy followed by surgical excision and then facial reconstruction along with skin graft was the preferred mode of treatment in this case.

**CONCLUSION**

Neurovascular hamartoma is a histopathological diagnosis made for small to medium-sized vessels and closely packed groups of well-formed nerve bundles with-in a loose connective tissue matrix and the inflammatory component is minimal or absent. These are relatively uncommon but with recently reported cases, it is definitely not as rare as previously considered. Neurovascular hamartomas have specific features, and therefore, they should not be considered as any reactive lesion or any hamartomatous lesions. Neurovascular hamartoma in fact represents a separate and unique entity.

**CLINICAL SIGNIFICANCE**

Neurovascular hematomas are relatively uncommon and have been very rarely reported in head and neck region. The rarity could be attributed to it being unrecognized and/or unreported. They constitute unique group of lesions with specific histopathological features. This case...
would be enhancing the knowledge of oral pathologist and maxillofacial surgeons. Therefore, this case would be apt in this upcoming journal.

REFERENCES