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Case Report

Nuchal Fibroma: A rare entity of neck masses

N. Alsaleh¹, H. Amanguno²

¹Department of Surgical Oncology, ²Department of Histopathology Kuwait Cancer Control Center, Kuwait

Abstract

A nuchal fibroma is a rare benign mass; it is a fibrolipoma-like soft tissue mass, usually located in the lower back and the neck. Males are affected far more often than females. It can occur extranuchally. It is a slow growing tumor and usually asymptomatic.

We present a rare case of a 39-year old female who was diagnosed with nuchal-type fibroma on final pathology. The proper management for this type of tumor is complete excision.

Keywords

nuchal, fibrolipoma, excision.

Introduction

Nuchal fibroma (collagenosis nuchae), is a benign soft tissue tumor that arises from the posterior cervical subcutaneous tissue (¹). This lesion can be easily misdiagnosed and under-reported in the literature. It is usually a slow-growing, asymptomatic solitary mass that occurs more frequently in middle-aged males. Histologically, it shows as a hypocellular dense collagen in the dermis and subcutaneous layer. Nuchal fibroma should be included in the differential diagnosis of soft tissue masses arising in the posterior neck, especially when the diagnosis is not definite (²).

We report an unusual case of a 39-year old female presenting with posterior neck mass for which optimal management would be careful total excision. This is performed for proper treatment and accurate diagnosis. We will also present a concise approach for posterior neck masses and the best way to evaluate a patient with a solitary neck mass.

Case Report

We present a 39-year old female with 10 year history of right posterior neck mass. It was gradually increasing in size causing social disturbance but no pain or history of infections. She is otherwise a healthy married lady with 3 children. Clinically, patient has a huge right sided posterior neck mass measuring 30x15cm, mobile and feels subcutaneous, not attached to underlying structures. No cervical lymphadenopathy (Figure 1). Patient was investigated with MRI scan of the neck which showed a soft tissue mass lesion in the right posterior neck region, multilobulated with thin capsule measuring 122x120x93mm. Cystic

Figure 1: Patient with posterior neck mass.

Corresponding author: Dr. Noha Al–Saleh, Department of Surgical Oncology, P.O Box142, Al–Jabreya, Postal Code 46302, Kuwait. Tel: 00965–25359339 Cell: 00965–99211999, Email: nohaalsaleh@yahoo.ca
changes with calcifications were noted. No infiltration of nodes or muscle was seen (Figure 2). Fine needle aspiration cytology of the mass was performed showing mainly blood cells and rare collagenous tissue with fibroblasts.

Patient was advised to undergo wide local excision of the mass under general anesthesia. The patient was placed in the left lateral position; elliptical skin incision was performed around the mass, flaps created all around it using sharp and blunt dissection. Muscle fibers were pushed away from the mass. The mass felt cystic in nature and a lot of feeding vessels were encountered during dissection, these were either clipped or cauterized. The mass was completely excised (Figure 3) and the skin was closed in layers. Patient did well post operatively; she had her drain out in day 3 post-op and was discharged home. Her skin clips were removed in post-op day 7 (Figure 4).

Gross examination of the specimen showed a soft tissue mass weighing 739gms measuring 15x12x8cm, cut sections showed a tan to grayish lobulated mass. Microscopic examination show haphazardly arranged bundles of collagen, with marked hypocellularity. There are areas of entrapped adipose tissue, blood vessels and nerve bundles. There was paucity of mitoses and necrosis. The mass had pushing border with peripheral areas of compressed fibrous stroma (Figure 5). Immunohistochemical stains showed positive staining with vimentin but negative for SMA, desmin, CD117 and ER. The diagnosis of nuchal type fibroma was made.

**Discussion**

Enzinger and Weiss first described Collagenosis Nuchae (CN) or nuchal fibroma (NF) in 1988. It is a rare benign soft tissue tumor that arises from the posterior cervical subcutaneous tissue with predilection for
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interscapular and paraspinal regions (3). There is no known etiology. It is a slow-growing, asymptomatic solitary mass that occurs more frequently in middle-aged males. Most tumors are about 3.5cm, but can be up to 8cm.

Histologically, it is characterized by hypocellular dense collagens in the dermis and subcutaneous layer with entrapped adipocytes and increased numbers of small nerves (4). The lesion that need to be differentiated from nuchal type fibroma (5) include sclerotic fibroma or circumscribed storiform collagenoma which is generally dermal lacks trapped adipose tissue.

The tumor cells are strongly positive for vimentin, CD34 and sometimes CD99. There is often (up to 2/3) a nuclear reaction with B–catenin(5).

Because of the benign clinical course and its close histopathologic similarity to other benign head and neck lesions, this lesion may be misdiagnosed and underreported. It was found that the mean age of patients affected by these tumors was 40 years old. There is a strong association with diabetes mellitus and also appears to be linked to Gardner’s syndrome (5), in fact it may be the initial manifestation of Gardner syndrome (6).

Nuchal fibroma should be included in the differential diagnosis of soft tissue masses arising in the posterior neck; other diagnoses include elastofibroma, fibrolipoma, desmoids–type fibromatoses and nuchal fibrocartilaginous pseudotumor.

Careful total excision is necessary for the treatment and accurate diagnosis. However, in patients with Gardner syndrome, up to 45% will develop desmoids–type fibromatosis at other sites, and so this should be searched for and excluded. Patients can develop a recurrence, so follow up is required (1).

Conclusion

Physicians are often asked to evaluate a patient with a solitary neck mass. Clinical evaluation of a solitary neck mass can be difficult because of the extensive differential diagnoses. These should include nuchal–type fibroma.

A nuchal–type fibroma is a rare proliferation involving the dermis and subcutaneous tissues, it is no longer called a nuchal fibroma, but instead a “nuchal–type fibroma” since it develops in other anatomic sites. These lesions are generally asymptomatic. We reported a case of a solitary posterior neck mass that was managed successfully by a simple excision. Follow up for these patients is very important in order to rule out recurrences.

References:
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