Case Report

Unusually Large Pilomatrixoma in the Neck

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Abstract

A 55-year-old Hispanic woman presented with a slow growing right-sided neck mass that was intermittently painful over the last year. There were no complaints of difficulty swallowing or breathing. Physical examination identified a four centimeter mobile, painless right submandibular neck mass. Computed tomography (CT) revealed a 3.1 cm lesion superficial to the platysma and fine needle aspiration was non-diagnostic. Excision of the mass confirmed a grey, well circumscribed mass with gritty consistency. Pathological examination revealed an epithelial neoplasm arranged in a nest of basaloid cells forming shadow cells among a multinucleate giant cell reaction. This neck mass was believed to be a pilomatrixoma, a rare, benign neoplasm arising from hair cortex cells. These masses most commonly arise in children, but can occur at any age in both males and females. Typically, pilomatrixomas are located on the eyebrows, scalp, trunk, and upper extremities. Symptoms typically include pain, tenderness, and inflammation from a solitary mass ranging from 0.5 to 3 centimeters. CT scan, ultrasound, and Magnetic Resonance Imaging (MRI) can all be used in the diagnosis, but fine needle aspiration is usually the test of choice. Pathology typically demonstrates ghost cells, basaloid cells, and calcium deposition. Treatment is complete surgical excision, and recurrence is as low as 1% following excision.

INTRODUCTION

Pilomatrixoma is a rare, benign neoplasm arising from hair cortex cells. These neoplasms were first described in 1880 as a skin tumor from sebaceous glands [1]. It is now known that pilomatrixomas are benign skin neoplasms that present as deep, subcutaneous lesions between the dermis and hypodermis [2]. Pilomatrixomas are also known as calcifying epitheliomas of Malherbe.

CASE PRESENTATION

A 55-year-old Hispanic woman presented with a slow growing right-sided neck mass that was intermittently painful over the past year. There were no complaints of difficulty swallowing or breathing, and no history of weight loss, fevers, chills, or night sweats. Physical examination identified a four centimeter mobile, painless right submandibular neck mass.

Computed tomographic imaging revealed a right-sided 3.1 cm lesion superficial to the platysma (Figure 1). Fine needle aspiration of the mass was non-diagnostic as the sample collected was largely acellular, except for a single group of epithelioid cells. Intraoperatively, the capsule of the mass was entirely superficial to the platysma. Circumferential dissection was achieved without any bleeding or spillage of mass contents. Excision of the mass confirmed a grey, well circumscribed, firm mass with gritty consistency. Pathological examination demonstrated an epithelial neoplasm arranged in a nest of basaloid cells undergoing abrupt trichilemmal-type keratinization forming shadow cells. A background of multinucleate giant cell reaction was present (Figure 2).

DISCUSSION

Pilomatrixomas typically present as slowly enlarging, poorly defined, non-ulcerated masses. The diagnosis can be made by fine-needle aspiration biopsy, which is the most common diagnostic technique. Histologically, pilomatrixomas are characterized by a nesting pattern of basaloid cells, accompanied by multinucleated giant cells and calcifications. The treatment of choice is complete surgical excision, and recurrence is rare, occurring in less than 1% of cases.

Figure 1 Preoperative CT imaging shows a well circumscribed soft tissue mass located in the subcutaneous tissue below the right angle of the mandible. The lesion is located superficial to the cervical fascia and measures approximately 3cm in diameter.
asymptomatic solitary masses in the head and neck region of children. However, these subcutaneous masses can arise at any age in both males and females. Pilomatrixomas are typically located on the eyebrows, scalp, trunk, and upper extremities [3,4]. Typical symptoms include pain, tenderness, inflammation, and can progress to abscess formation, ulceration, or bullous lesions. Typically they present in an area of previous trauma [2]. Commonly, pilomatrixomas are clinically misdiagnosed as dermoid, trichilemmal, or sebaceous cysts, parotid or giant cell tumors, or lymphadenopathy [4].

Grossly, the appearance of these masses can range in color from normal to white to blue [5]. The size of pilomatrixomas typically can range from 0.5 to 3 centimeters [6]. One study found that the peak age of presentation was between 5 to 15 years, with a second peak arising between 50 and 65 years of age. This same case review analyzed 209 cases of pilomatrixoma, with size ranging from 0.5cm to 6cm [7]. However, 37% of these cases were between 1.0 and 1.5cm. These masses usually are solitary, but in 2% to 3% of cases they can present as multiple tumors [6]. Pilomatrixomas can also present with the “tent sign” which occurs due to the depression of one side of the lesion that causes elevation of the other side [4]. Upon computed tomographic examination, pilomatrixomas appear as radiopaque subcutaneous lesions with clearly delineated margins and calcifications [4]. On ultrasound examination, pilomatrixomas show round, demarcated hyperechogenic masses with a dense posterior acoustic shadow [4]. Ultrasound can be especially helpful when diagnosing pilomatrixomas in children. MRI examination demonstrates heterogeneous signal intensity with areas of delayed enhancement and can help determine the depth of the mass [8]. Since this condition can be viewed as a diagnosis of exclusion, many physicians will also obtain other tests, such as fine needle aspiration, to rule out malignancies.

To obtain a definitive diagnosis, fine needle aspiration is usually the test of choice. Pathological examination usually will show the presence of ghost cells, basaloid cells, and calcium depositions [8]. Ghost cells, also known as shadow cells, are enlarged eosinophilic epithelial cells devoid of a nucleus and can be found in solid sheets. There can also be the presence of a foreign body reaction with giant cells [6].

Interestingly, high levels of beta-catenin are implicated in pilomatrixomas. Beta-catenin normally links E-cadherin to the actin cytoskeleton, and is normally found in the hair follicle matrix. Accumulated mutations in beta-catenin translocate into the nucleus and activate the transcription of Lef-1, thus resulting in abnormal cell proliferation and ultimately a pilomatrixoma [9]. Further, beta-catenin is regulated and destroyed by the tumor suppressor APC gene. As a result, pilomatrixomas are associated with Gardner syndrome. Other conditions associated with this lesion are Steinert disease, myotonic dystrophy, and sarcoidosis [8].

When considering the possibility of pilomatrixoma, the differential diagnosis should include epidermal cysts, dermoid cysts, follicular cysts, steatocystoma simplex, cylindroma, and metastatic carcinoma [6,8]. The treatment of pilomatrixoma is complete surgical excision with possible removal of the overlying skin due to the lesion adhering to the dermis [8]. Recurrence of pilomatrixomas following excision is as low as 1% following complete excision [4].

Given the fact that most pilomatrixomas occur in children, and are usually less than 3cm in size, we find this case to be particularly interesting. Although the diagnosis of pilomatrixoma is not common, we believe that it is important to consider both the diagnosis and histology when evaluating an enlarged neck mass in both children and adults.

REFERENCES


Cite this article