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PILOMATRIXOMA
(A CASE REPORT AND REVIEW OF LITERATURE)

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SUMMARY: Pilatrixoma is one of the hair matrix cells. More than 60 % of the tumors appear in the first two decades of life and most commonly involve head, neck and upper extremities. These tumors are occasionaly confused with other neaplasms. A 2,5 yearl old white boy who had a right supraclavicular pilomatrixomas is presented here and discussed under the scope of the recent literature.

Pilomatrixoma was first described in 1880 by Malherbe and Chenantais and there after acquired the name calcifying epithelioma of Malherbe (2,5,9,10,12). They speculated that the origin of this tumor was sebaceous gland. In 1961 the name pilotamtrixoma was proposed and evidence supporting the genesis of this tumor from primitive germ cells which differantiated toward the formation of hair matrix cell (2,3,9). Pilomatrixoma is a benign skin neoplasms nearly always less than 3 cm. in diameter and it usually apperas as a slow-growing solitary, asymptomatic dermal or subcutaneous mass. It is usually seen within the firs two decades of life and most commonly involved the head and neck . (52 % of cases) (1-3,9,12). Treatment of this tumor is excisional surgery. This procdeure is generally curative and recurrences are rare (2,4,5,12). Surface tumors are quite rare in childhood. Since they look primary or metastatic lymphadenopaties due to their localization, their differential diagnosis assumes importance. Therefore, aspiration or excissional biopsy must be performed for accurate diganosis. A pilomatrixoma case is presented here which was seen at our Pediatric Hematology-Oncology outpatient clinic and which was impossible to diagnose clinically. A pathological diagnosis was made.

CASE REPORT

A 2,5 year old white boy presented with 2 years history of slowly growing asymptomatic nodule in the right supraclavicular region. In the last 3 months the tumor had grown rapidly. A 1,5 x 2 x 1 cm. firm well-circumscribed, and mobile nodule was noted in the subcutaneous tissue of the right supraclavicular region. The overlying skin showed bluish discoloration. There was no local lymphadenopathy. The liver and spleen were nonpalpable. Other physical and laboratory (hematological, biochemical and radiological studies) findings were normal. Due to suspected malignant neoplasm. the nodule was extirpated. Pathological diagnosis was pilomatrixoma. Histopathologic findings: (Fig. 1-3) An excissional biopsy procedure produced 1,5 x 2 x 1 cm. grayish-brown elastic material. Microscopic examination showed; pseudoencapsulated masses composed of basaloid cells or shadow cells. Around and among these cells, central keratinizing cells or keratin pearls, and focal calcifications were noted. Foreign body giant cells admixed with dense inflammatory infiltrate were also seen.

DISCUSSION

Pilomatrixoma is a rare benign tumor at hair matrix-cells usually seen in the younger

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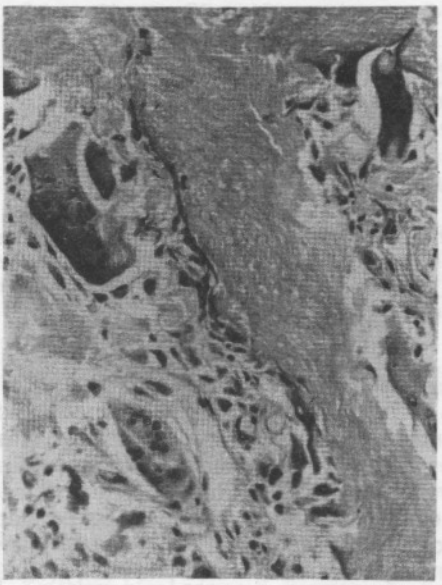

Figure 1: Anucleated shadow cell with foreing body giant cells (hematoxylin-eosin x 310)

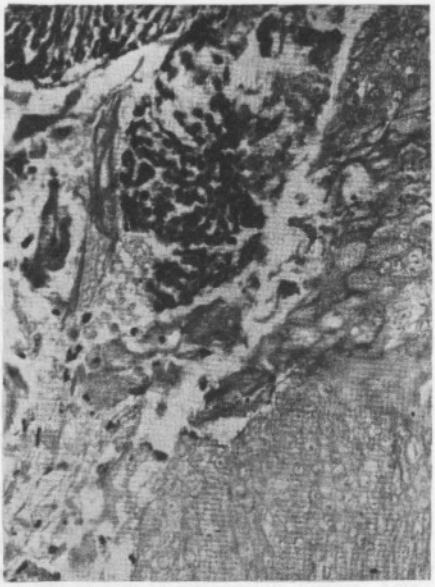

Figure 2: Anucleated shadow cells and ısland of dark basaloid cells (hemotdxylin-eosin x 310)

age groups (1,2,4,10,12). Approximately 1 in 2000 biopsy materials are diagnosed (5). Forty per cent of all cases are younger than 10 years of age and 60 % of all cases are younger then 20 (7,9,12). The mast common localization of the tumor is the head and neck, wheras upper exterimities, trunk, lower extremities and scalp can be involved as well (2,9,12). Generally, it is solitary, slow growing well encapsulated and is smaller than 3 cm. in diameter. However Sasani et al. showed that it can be aggressive and can metastize an relapse (3,4,6,9,11). The age, localization, size and character of our case patient were all in accordance with the literature.

Aspiration biopsy is inappropriate for the diagnosis of pilomatrixoma (1,9,12). Histopathological differential diagnosis sholud be made among sebaceous and tri-

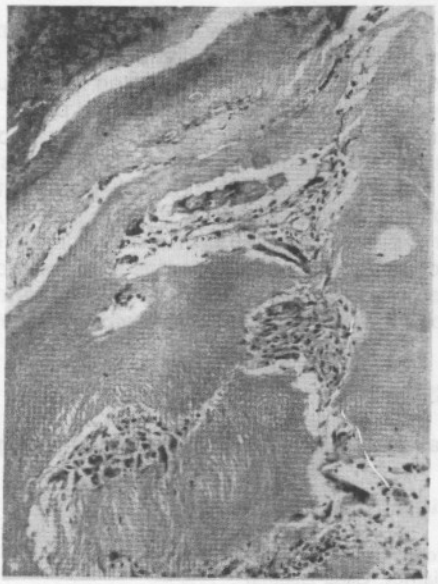

Figure 3: Anucleated shadow cells, focal calcification and foreign body giant cells (hematoxylin-eosin x 125)

cholemmal cysts, granulomathous lymphadenopathies, squamous and basal cell carcinomas (6-9). The tumor is composed of basophilic and shadow cells which have come together in irregular epithelial islets. Basophilic cells dominate in new tumors but they tend to decrease in later stages. Keratinizaiton centers and melanocytes can be seen among them. Sometimes, immature hairs, small sebaceous gland, and trichohyalen granules can be detected. In old tumors, calcifications are seen. Another finding, specific to pilomatrixoma is the perecence of shadow cells whose centers don't stain. These cells can not be seen in basal and squamous cell carcinomas. Basophilic cells are arranged in palisadic rows, which helps to differantiate pilomatrixoma from tricholemmal masses. Prominent nucleoli of the cells differantiate pilomatrixoma from basal cell carcinoma (1,7,12). Lymphadenopathies should be considered in differantial diagnosis (12). Hematological and biochemical tests and clinical findings can be helpful but the definite diagnosis is pathological. There fore the tumor should be excised completely. Excision is sufficient in most cases but if the tumor relapses, wide resections and radiotherapy may be needed (4,9). In our case, after the necessary preliminary test were completed, complete excision was performed. The patient has been fallowed up 3,5 years until without any sign of illness.

Pilomatrixoma should be considered in childhood solitary subcutaneous nodules, progressing slowly and giving no specific clinical or laboratory findings. once the diagnosis is made, surgery should be the choice of therapy.

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