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RESEARCH ARTICLE

PRIMARY CUTANEOUS MUCINOUS CARCINOMA OF ANTERIOR CHEST WALL: A RARE ENTITY

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ABSTRACT

Primary cutaneous mucinous carcinoma or Primary cutaneous colloid carcinoma is a rare malignant tumor of the eccrine glands. The tumor may be asymptomatic overall several years but generally it presents as a single hard swelling with a smooth surface, sometimes ulcerated or crusted. The tumor mainly affects middle aged man more than females. The tumor is resistant to radiotherapy and chemotherapy, so the treatment of choice is wide local excision. Here we present a case of primary mucinous carcinoma of anterior chest wall which is a very rare site of presentation.

INTRODUCTION

Primary cutaneous mucinous carcinoma or Primary cutaneous colloid carcinoma is a rare malignant tumor of the eccrine glands.¹ It was first documented by Lennox *et al.* in 1952, and was first designated by Mendoza and Helwig in 1971. Clinically the tumor presents as a single hard swelling with a smooth surface, sometimes ulcerated or crusted. Swelling may be asymptomatic over several months or even years.^{3,4} The estimated incidence of cases is 0.07 per million person years mainly affecting middle aged males than females. The commonest site of primary cutaneous mucinous carcinoma is head and neck- eyelids (50% cases), followed by face (excluding periorbital area (19.5%) and scalp (17%). Whereas the axilla, trunk, lower extremities and genital areas are the other uncommon sites.² Current report presents a case of primary cutaneous mucinous carcinoma of swelling anterior chest wall in a 66 years old male.

CASE REPORT

A 66 years old Indian man presented to the surgical out-patient department of our hospital with swelling over anterior chest wall.

Apparently, the swelling had been present since 2 and half months which was not increasing in size. There were no other complaints. On examination, an immobile, hard swelling measuring 2X3cm was noted over anterior chest wall. Excision biopsy was done and the specimen was sent for histopathological examination. Gross examination revealed multiple grey brown to tan soft tissue pieces measuring 2.0 X0.5X 0.2 cm in size and partially gelatinous in consistency. Microscopically, the sections revealed a dermal tumor composed of large pools of mucin separated by thin fibrovascular septa and floating tumor cells arranged in nests, glands, and cribriform patterns. The individual tumor cells were columnar, had a hyperchromatic nucleus and moderate amount of cytoplasm (Figure 1). The mucin was periodic acid Schiff (PAS) positive (Figure 2). On immunohistochemistry, the tumor cells showed positivity for CK 7 (Figure 3), CEA (Figure 4) and EMA (Figure 5) while these were negative for CK20, TTF-1, SMA and CK5/6. The clinical investigation and imaging studies demonstrated the absence of regional lymph nodes or distant metastases and ruled out a primary cancer at any other site, and it was concluded that the tumor was primary. Thus, the lesion was reported as primary mucinous carcinoma of skin.

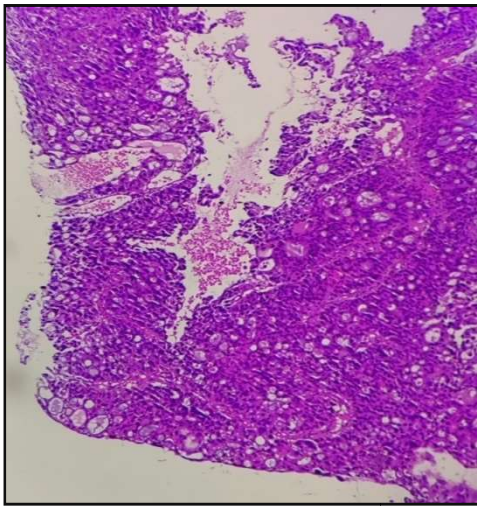


Figure 1. H & E section showing large pools of mucin separated by thin fibrovascular septa and floating tumor cells arranged in nests, glands, and cribriform patterns (10X)

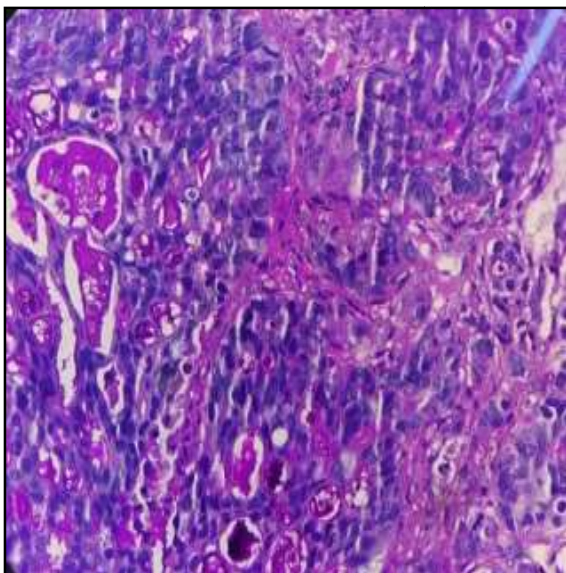


Figure 2. Mucin which is PAS positive (40X)

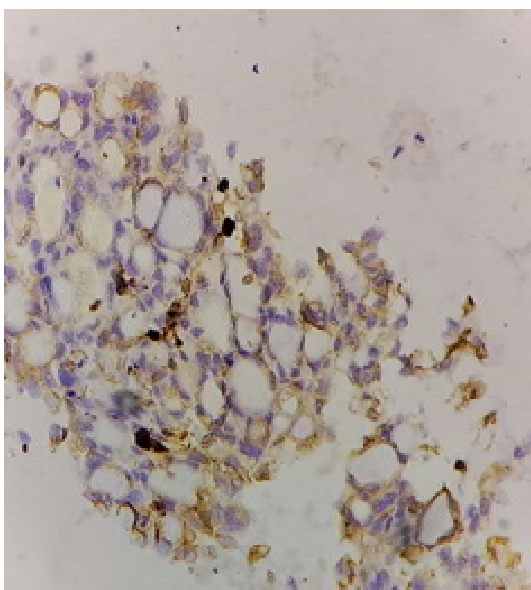


Figure 3. CK7 Positivity in tumor cells

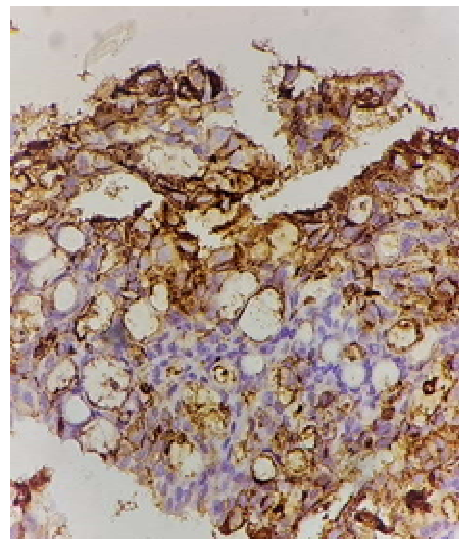


Figure 4. CEA positivity in tumor cells

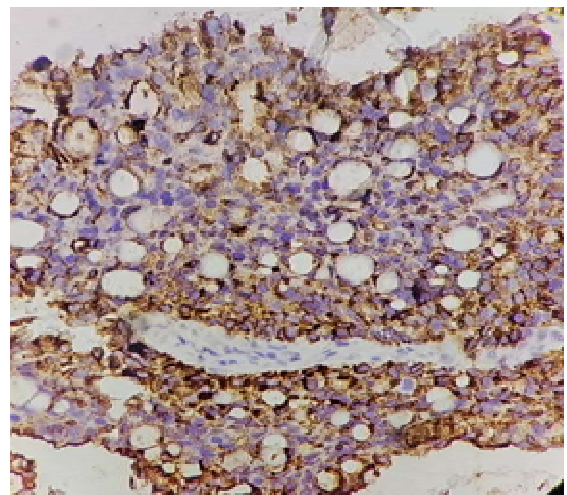


Figure 5. EMA positivity in tumor cells

DISCUSSION

Primary cutaneous mucinous carcinoma is a rare low-grade malignant neoplasm involving the adnexa. This tumor generally affects the individuals between fifth to seventh decades of life with increased predominance in males than females.² It occurs commonly in the head and neck region and lesions typically present as erythematous, asymptomatic nodules measuring 0.5 to 7 cm in diameter.⁴ It is thought to arise from sweat glands. However, it is still controversial whether this neoplasm has an eccrine or apocrine origin.³ The mucin in primary cutaneous mucinous carcinoma is sialomucin which is PAS positive, diastase resistant and also shows positive staining with mucicarmine, colloidal iron and alcian blue at pH 2.5. Electron microscopy shows two types of cells-dark cells and light cells. Dark cells are responsible for secretion of sialomucin.

Primary cutaneous mucinous carcinoma shows positivity for CK7, CAM 5.2, CEA, EMA, GCDFP-15, human milk factor globulins, S100, alpha-lactalbumin. The tumor is found to CK7 positive in 100% cases, while p63 and CK5/6 positivity was reported in 40% and 20% cases respectively. Strong ER positivity and variable PR positivity has been observed in the lesions. It is very rare to be skin as primary origin of carcinoma because the majority of the skin lesions are actually metastatic. The metastatic lesions from the breast or colon are most likely to mimic mucinous carcinoma of the skin. So, it can be very difficult to differentiate the primary mucinous carcinoma from metastatic tumors from these sites.

The metastatic colorectal mucinous carcinoma may be excluded by the absence of expression of CK20.¹ Metastasis from breast primary is even more difficult to exclude, however a positive immunostain for myoepithelial markers i.e, p63 and CK5/6 can be helpful to exclude metastatic breast mucinous carcinomas.² As this tumor is resistant to radiotherapy and chemotherapy, the treatment of choice is surgical. Wide local excision is done with margin of 1 cm. The prognosis is good however there is tendency for late recurrences and rare metastasis.⁴ Long-term follow-up with regular clinical investigation to detect tumor recurrence and metastasis is mandatory.

CONCLUSION

Primary cutaneous mucinous carcinoma is a rare neoplasm of skin occurring in middle aged males with a good prognosis, which is usually managed by wide local excision.

Histopathology plays a very crucial and important role in diagnosing this entity. The lesion can be categorized further by using immunohistochemistry.

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