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"RARE TUMOUR OF THE HAND: MALIGNANT ACROSPIROMA OF THE HAND"

Pathology

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ABSTRACT

Background: Malignant acrospiroma is among one of the rare tumors of the eccrine sweat glands. It accounts for around 6% of all malignant eccrine tumors. It typically presents as large ulcerated nodules, and diagnosis can be challenging as it has great overlap with its benign counterpart. There are limited cases reported for hand acrospiroma's.

Case presentation: We report a case of malignant acrospiroma presenting as swelling which was treated with surgical excision. The patient was referred to oncosurgery for chemoradiaton.

Conclusion: Wide surgical excision, with or without prophylactic lymph node dissection, is the treatment of choice. Evidence is lacking on the efficacy and/or advised regimen of chemoradiotherapy. Prognosis is poor with systemic disease. More studies are needed for the management of this disease.

KEYWORDS

Acrospiroma, Malignant.

BACKGROUND

Malignant acrospiromas arise from the intradermal duct of eccrine sweat glands. These tumours account for nearly 6% of malignant eccrine tumors [1] compared to their more prevalent benign counterparts [2]. They are also called as clear cell hidradeno carcinoma, malignant clear cell hidradenoma, and malignant hidradenoma [3, 4]. Disease usually manifests in middle-age around 40-50 years of age [5]. Some reports show disease is more common in females [6], but there does not seem to be obvious gender predominance [4, 7, 8]. These lesions present as slow growing nodules that ulcerate and start draining [6, 9]. They range in size from 0.5 to 10 cm [6, 9]. They usually involve the head, neck, and extremities; less common sites include the chest and breasts [1, 4, 5, 9, 10]. These tumor's are very aggressive having more than 50% local recurrence rates [11]. Wide local excision is the treatment of choice [12–14]. The efficacy of adjuvant chemotherapy is controversial. Adjuvant radiotherapy has been shown successful in some cases [4, 12, 13].

CASE PRESENTATION

A 23 years male patient presented with a recurrent swelling of right forearm near wrist flexor aspect. Patient had history of excision of a soft tissue swelling of approximately 3x2 cm size 3 years back by a local doctor, HPE was not done at that time. Patient presented to our department with recurrence of swelling at same scar line. Swelling slowly progressed to size of 4x3 cm. It was non-tender, non- fluctuant and firm in nature. Skin overlying the swelling was free expect at previous suture line where it was tethered. There was no neurovascular deficit. After written informed consent the patient was taken up for exploration and excision of swelling under regional anesthesia and tourniquet control was done. Previous scar was excised, and swelling was dissected out, swelling was found to lie in subcutaneous plane and was surrounding the median nerve and nearby tendons, these structures were preserved. The specimen was taken out and sent for HPE.

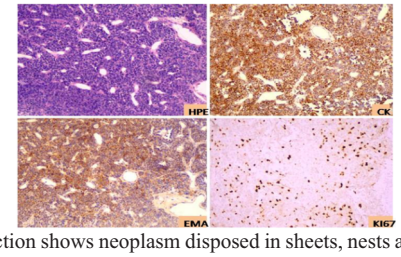

Figure:Section shows neoplasm disposed in sheets, nests and lobules of cells admixed with variable sized thin walled blood vessels. Variable size small and large ductal lumina are lined by cuboidal cells. Individual tumor cells are round to oval, moderately pleomorphic nuclei with mild tomoderate cytoplasm.Fair number of mitotic figures are also seen [H&E stain,x 400].Cytokeratin diffuse positive and Vimentin focal positive in tumor cells. S-100& Chromogranin negative [IHC Stain,x 400].

Patient recovered well with no neurovascular deficit.

DISCUSSION

Malignant acrospiroma is one of the rare tumor's of the eccrine sweat glands. It can arise de novo or transform from a previous benign lesion. Our patient did not have any history of prior malignancy.

Wide local excision is the treatment of choice [12]. Wildemore had compared outcomes of Mohs micrographic surgery to conventional surgery; all patients who underwent Mohs micrographic surgery had no local recurrence [14].

Our patient was treated with surgical excision. Even with wide surgical excision, recurrence rates as high as 50% have been documented in studies [15]. As the lesion has an aggressive nature and there is a tendency for lymphatic invasion, some authors have advocated for prophylactic lymph node dissection [16]. There is little evidence on the efficacy of adjuvant chemotherapy [17].

Malignant acrospiroma is one of the rare tumor's that originates from the eccrine sweat glands. Aggressive in its nature, its diagnosis is challenging, and specific tumor markers and gene mutations are not defined.

Wide local excision is the treatment of choice. There is lack of evidence on the efficacy and/or advised regimen of chemoradiotherapy. Prognosis is poor with systemic disease. More studies are needed for the management of this disease.

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