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Epithelioid Sarcoma of the Perianal Region - A Case Report

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Abstract

Aim: To highlight a rare case of epithelioid sarcoma of the perianal region

Case Report: A 36 year old male with no known co-morbidities , came with complaints of swelling with pain in the right perianal region for the past 1 year ; associated with on-off purulent discharge .On examination, a 5 x 3 cm firm-hard swelling (4m from anal verge) was found in the right perianal region; warmth and tenderness present; pus pointing over the swelling present. A clinical diagnosis of Chronic perianal abscess or Antibioma was made and patient was taken up for Incision and Drainage. Intra-operatively a hard mass was found in the right gluteal region from which biopsy was taken.HPE was reported as Epithelioid Sarcoma. Hence, wide local excision of tumour was done with adequate margin of clearance followed by post-operative radiotherapy.

Conclusion: The Epithelioid sarcoma(ES) is a rare, high-grade malignancy, with known propensity for local recurrence, regional lymph node involvement, and distant metastases. Perineural and vascular invasion can also be observed. This tumour is often mistaken for chronic inflammatory processes, necrotising granulomas, and various fibrohistiocytic tumours. Aggressive treatment with preoperative or postoperative radiation therapy combined with wide local excision has to done once diagnosis is established via histopathological confirmation (Vimentin and EMA positive). Because of its potential for aggressive behaviour, clinicians must be aware of the presenting behaviour of ES in order to avoid misdiagnosis.

Keywords: Epithelioid Sarcoma , Perineal Sarcoma

Introduction

Epithelioid sarcoma (ES) ,the most common primary soft tissue sarcoma of the hand, is a rare, high-grade malignancy. Incidence – 1 in 10,00,000. It occurs in all races. It usually affects adolescents and young adults. Extremities are commonly affected. In 1961, Laskowski first identified it as "sarcoma aponeuroticum" because of its association with aponeurosis and surrounding structures[1]. Less than a decade later, when

recategorizing 62 previously misdiagnosed tumours, Enzinger coined the new word "epithelioid sarcoma"[2]. This tumour has also been mistaken for chronic inflammatory processes, necrotizing granulomas, and multiple fibrohistiocytic tumours due to its epithelial and mesenchymal differentiation[3]. Clinically and histologically, these lesions were once documented as malignant and nonmalignant conditions[2]. In young men, ES often occurs in a banal manner, but in addition to regional lymph node involvement and distant metastases, it is inherently prone to local recurrence, hence , is of great significance[4-8]. Misdiagnosis of this tumour can result in delayed and improper treatment, adversely affecting the survival of patients. **Case Report** A 36 year old male, with no known co-morbidities, came with complaints of swelling and dull-aching pain in the right perianal region for the past 1 year; associated with on-off purulent discharge.On examination , a 5x3cm hard swelling was found on the perianal region (4cm from the anal verge) on the right side.Warmth and tenderness present over the swelling. Pus pointing over the swelling was noted. There were no palpable inguinal nodes. A clinical diagnosis of a Chronic perineal abscess or an Antibioma was made and the patient was taken up for Incision and Drainage under regional anaesthesia. Intra-operatively, about 5ml of us was drained , following which, a hard mass was felt in base of the cavity. A biopsy was take from this site and sent for histopathological examination(HPE). The biopsy was reported as an Epithelioid Sarcoma. Hence , metastatic work up was done ; MRI pelvis was done; HRCT lung was done to rule out metastasis. Wide local excision of the tumour was done with an adequate margin of clearance , followed by post-operative radiotherapy.

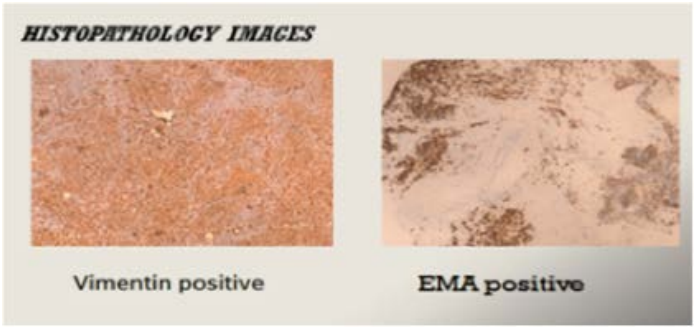

Figure 1: Histopathology Images

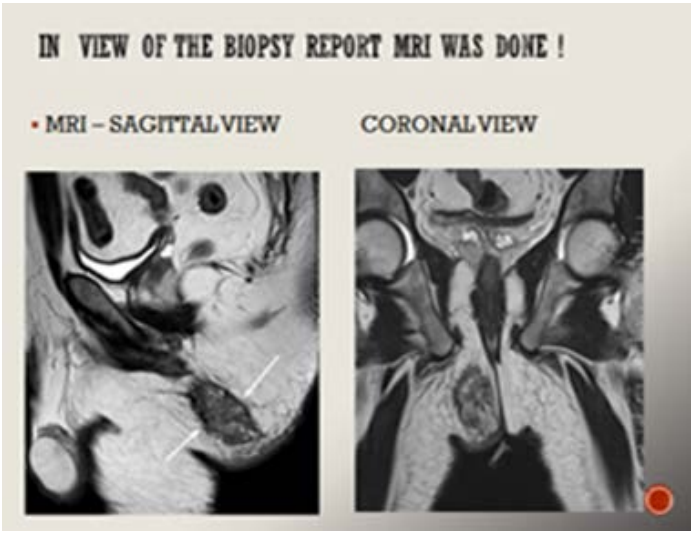

Figure 2 : MRI Pelvis

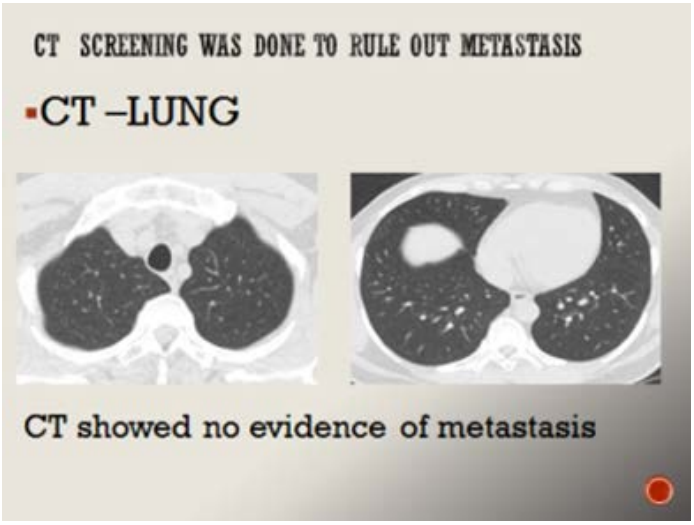

Figure 3 : HRCT Lung

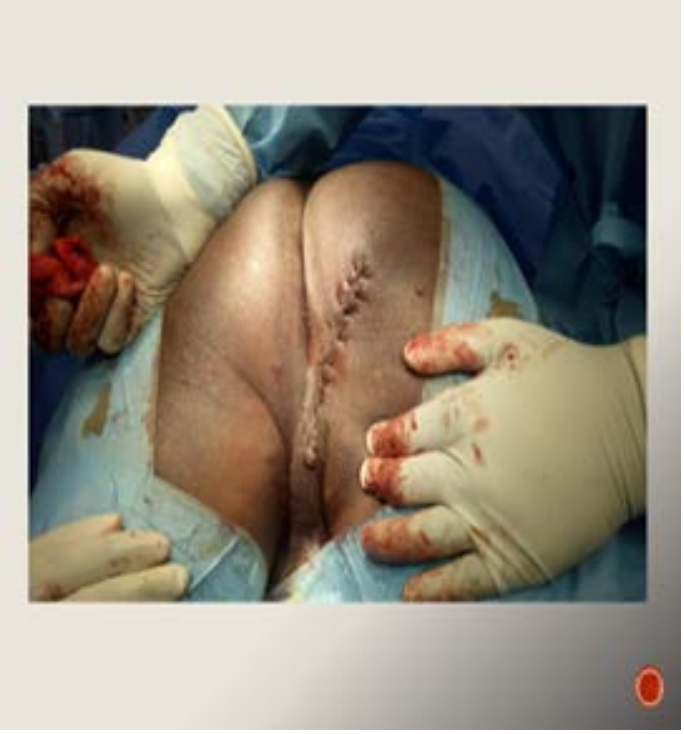

Figure 4 : Post-operative Picture

Discussion

On physical examination, ES is described as a firm, non-tender, slow-growing tumour with a predilection for the hands, fingers, and forearms[4,8-10]. ES initially appears as a single nodule. However, multiple nodules representing local disease spread may be present at the time of diagnosis[7]. Tan-white, nonencapsulated nodules with infiltrating borders are the most prevalent gross appearance[2]. It has been noted that trauma precedes tumour growth. ES affects young adults predominantly in their second or third decade of life, but may occur at any age. With a ratio approaching 2:1, males are disproportionately affected[2,5,8-13].

ES may be present for months to years before the patient seeks medical attention because of it's tendency toward non-painful, indolent growth[2,5-14]. If other clinical factors point towards malignancy, this "benign nature" should not prevent the clinician from performing a biopsy. Occasionally, there may be pain and tenderness, drainage, contractions, muscle weakness, numbness, and tingling[5,9,11,12]. Function limitations are usually not observed. By the time a patient seeks medical care, ES may be multifocal. The tumour may have spread proximally throughout the tendons, fascial planes, and aponeurosis, resulting in nodular or pseudogranulomatous growth. The secondary nodules can be solitary or multiple and range in size from a few millimetres to several inches[2,4,5,9,12]. They are often subjected to central necrosis, bleeding, and ulceration[2,4,5,8,9,11]. Periosteal bone invasion can occur[2,14]. The scalp, orbit, parotid gland, palate, penis, perineum, vulva, and buttocks are other areas where the tumour has been reported[17-22]. A common feature of ES is dissemination through the sub-dermal lymphatic vessels or the blood stream. When distant metastasis occurs, the tumour can involve one or more lymph nodes and these diseased nodes can enlarge up to several centimetres[2,5,8,9,12]. While a higher rate of lymph node involvement has been shown in adult patients with ES than in other soft tissue tumours, this trend is less evident in paediatric patients[13]. The most frequent site of distant organ metastasis is the lungs[2,5,7,9]. Through histopathological examination, the diagnosis of ES is made. Some clinicians may choose to receive imaging prior to having a subcutaneous tumour biopsy to evaluate for subclinical involvement and the best means of biopsy. The chosen modality for imaging is magnetic resonance imaging (MRI), as opposed to computed tomography scans and X-rays, due to its ability to visualise soft tissue information[10]. MRI can also be used after excision to distinguish the residual tumour from postoperative disease[10,23]. The three ES variants are epithelioid, spindled, and mixed, with epithelioid as the main type. These cells are large, circular, oval, or polygonal, with a clear or vesicular, centrally positioned nucleus and abundant, deeply acidophilic cytoplasm[2,5,12]. Well-defined cytoplasm and distinctive

cell boundaries are found in the cells. Almost always, mitotic activity is reported. When ES originates in the superficial dermis and subcutaneous tissue, ulceration can occur. It can be attached to tendons, tendon sheaths, or fascia as ES stems from under the fascia[8,11]. ES tends to be larger and deeper than the superficial forms located on proximal portions of the body[9,12]. ES also exhibits perineural and vascular invasion. The course of ES is often unpredictable, and patients often present with extensive disease, lymph node metastases, or distant metastases. A local recurrence rate of 85% and a distant metastatic rate of 30 percent were demonstrated by Enzinger's initial report on ES[2]. A metastatic rate approaching 50 percent was reported by more recent authors[5,8,9,11,26]. The lungs are the principal site of metastatic disease, as is true for most soft tissue sarcomas[2,5,7-9,10]. Metastasization of ES to the lymph nodes, skin, scalp, brain, digestive tract, liver, kidneys, and musculoskeletal system has also been documented[11]. Local recurrence often occurs within 1 to 2 years of treatment, and distant metastases often continue to develop in these patients[5,8]. It was found by Spillane et al that both local recurrence and regional nodal metastases resulted in increased distant metastatic disease, thereby reducing overall survival[8]. It was reported that the median postmetastatic survival was eight months[8].Chase and Enzinger reported that the tumour width was directly proportional to the rate of metastasis and that the tumour width was directly correlated with a lower survival period of 10 years[9]. The same findings were demonstrated by numerous other case series[5,7,9,10,12-14,16]. Interestingly, a positive prognosis has been associated with female gender, but patient age does not seem to alter survival at the time of diagnosis.There have been inconsistent reports of greater tumour depth, vascular invasion, and necrosis to indicate a poorer prognosis[11,12,16]. ES's anatomical location seems to play a role in prognostication. In lesions proximal to the elbow or knee, the overall survival and metastases-free survival are worse[7,15].This can be explained by the fact that "proximal-type" epithelioid sarcomas tend to have unfavourable characteristics, such as increased width, location of the deep-seated tumour, and preferential pelvic, perineal, and genital region involvement[7,13]. Conversely, a more favourable result is predicted by location on the distal extremities[13]. The primary treatment for patients with ES is radical tumour excision. It is necessary to obtain negative tumour margins, but optimal function, particularly of the upper limb, remains a potential goal[2,5,8-11,13,16]. Due to the high rate of local recurrence and distant metastases, marginal excision is inadequate. Data does not appear to support routine sentinel lymph node biopsy (SLNB), although it is sometimes advocated because of the high rate of regional recurrence. It is also thought that SLNB identifies people who may benefit from systemic therapy[7,10]. When metastases of the lymph nodes are present, therapeutic lymph node dissection is indicated. ibHalling et al suggest that surgical resection of single or small numbers of metastatic lesions may result in increased long-term survival in the case of in-transit metastases. Amputation may be necessitated by local recurrence[14]. While the effectiveness of radiation therapy as an adjunct to surgery has been clearly demonstrated in soft tissue sarcomas, due to small sample size and limited follow-up, studies of effectiveness in ES are limited[27,28]. With the addition of preoperative or postoperative local radiation therapy when combined with radical surgery, a decreased incidence of local recurrence has been reported. For patients with marginal primary resection, local regional recurrence, or palliative treatment, radiation can also be considered[8]. In the case

of metastatic disease, adjuvant chemotherapy appears to be indicated, but in nonmetastatic ES, this is less evident[26]. Additional long-term follow-up studies would be beneficial in further addressing the role of adjuvant therapy in treating ES. Limited experience with isolated limb perfusion demonstrates that multifocal or large unresectable tumours may be beneficial, enabling tumour size reduction with subsequent radical excision[7,8]

Conclusion

The Epithelioid sarcoma(ES) is a rare, high-grade malignancy, with known propensity for local recurrence, regional lymph node involvement, and distant metastases. Perineural and vascular invasion has also be observed. Chronic inflammatory processes, necrotizing granulomas, and different fibrohistiocytic tumours are often mistaken for this tumour. Aggressive treatment with preoperative or postoperative radiation therapy combined with wide local excision has to done once diagnosis is established via histopathological confirmation(Vimentin and EMA positive)[5,7] . Due to its potential for aggressive behaviour, in order to prevent misdiagnosis, clinicians must be aware of the presenting behaviour of ES.

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