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## **CYSTIC FOREIGN BODY GIANT CELL GRANULOMA IN XERODERMA PIGMENTOSUM**

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### **Abstract**

A case of cystic foreign body giant cell granuloma is presented. The patient, nineteen years of age, known case of Xeroderma Pigmentosum, presented to the maxillofacial unite, Basrah General Hospital, Iraq, with slowly enlarging cervical cystic mass. Examination revealed cystic swelling in the mid-left side of the neck of six months duration. The mass was not tender and was immobile. Aspiration revealed straw colored fluid. The excised mass, showed cystic lesion, the base of which had an extensive soft tissue growth, histologically consisted of foreign body giant cell granulomas. There was no recurrence at a follow-up of 17 years.

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### **Introduction**

Xeroderma Pigmentosum is a rare hereditary disease transmitted as an autosomal recessive disorder, characterized by extreme sensitivity to sunlight<sup>1</sup>. Patients develop multiple skin and mucosal cancers in the sun light exposed areas often within the first few years of life. There is a deficiency of the enzyme required for the nucleotide excision repair of the ultra-violet light induced mutations. Most persons with Xeroderma Pigmentosum appear normal at birth but within time, an exorable destruction of skin sets in, freckles and solar keratosis usually have formed by late infancy. In minor proportion of affected persons, neurological abnormalities including mental deficiencies are an integral part of the clinical syndrome. Many others suffer serious corneal damage, the consequence being varying degrees of blindness. Angiomas, basal cell carcinomas and squamous cell carcinoma are among the skin tumors. Oral squamous cell carcinomas are frequently found as well<sup>2,3</sup>.

The Author had an experience with one family of 13 children, 9 girls and 4 boys, of them four girls were involved with Xeroderma Pigmentosum. A total of 32 operations were conducted by the same surgeon (author) for excision of 7 Angiomas, 7 Basal cell carcinomas, and 18 squamous cell carcinomas .

Follow-up for 17 years, none of the patients had recurrence at the original primary site, neither had shown metastasis to cervical lymph nodes.

In one case, the patient at the age of nineteen years developed a very rare condition of cystic foreign body granuloma in the neck; this has not been described before. The author presents the first such case.

### **Case Report**

A 19 year old female patient, a known case of Xeroderma Pigmentosum, presented with a non-painful cystic mass at the mid-left side of the neck. The patient has noticed a small soft swelling six months earlier, since then it slowly increased in size. The patient has nine

sisters and three brothers. Three sisters are involved with the disease. The father and mother are close relatives. None of them or their parents have the disease neither they recall the occurrence of the disease in the family. The patient has nine operations conducted by the same surgeon (author), the first when she was two years old, and the last when she was thirteen years. The operations were to excise six squamous cell carcinomas involving the anterior part of the tongue, the vermilion border of the lower lip,

commeasure of the mouth, skin of the face and the scalp; these were presented as erosive or, ulcerative lesions and exophytic lobulated partly infiltrative masses (fig.1). Other tumors were basal cell carcinoma involving the infraorbital region, skin of the nose; others were angiomas involving the medial canthal area and lower eyelid. The patient did not recall any trauma to the face or neck neither she had any previous surgery to the neck.

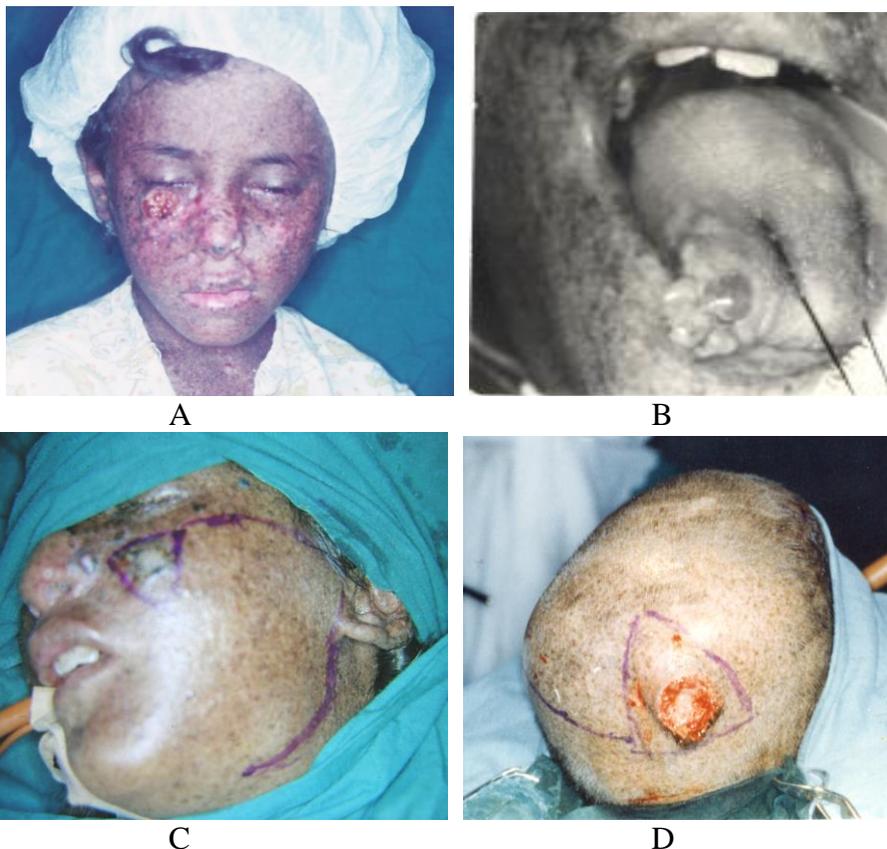


Fig 1: Shows the patient with Squamous Cell Carcinoma with different clinical presentations in multiple sites in different ages. A, at the age of 6 years, an ulcer involving the infraorbital region. B, at the age of 7 years, showing an exophytic lobulated mass in the tongue. At the age of 12 years, shows a nodular mass with indurated base. D, at the age of 13, there was an exophytic mass with ulcerative surface in the scalp.

On examination, Apart from her skin disease, she is healthy looking, no signs and symptoms of any other systemic disease, no neurological abnormalities. There is mild corneal damage. The patient had scars in the face and lips from previous operations. There is cystic swelling of 6x6 cm in diameter situated in

the mid-left side of the neck, not tender, immobile in either direction, the base of which seems to be indurated and fixed to the underlying muscles, the covering skin is normal in color and texture, (fig 2 A, B). Aspiration of the cystic content revealed clear yellowish to brown color fluid (fig 2 C).



Fig.2: A-The patient at presentation shows cystic mass at the mid-left side of the neck. B-aspiration revealed straw colored fluid. C-7 days Post-operative. D-12 years post-operative.

All hematological investigations were within normal limits, clear chest X-ray. Seventeen years follow-up for this family did not show metastasis to cervical lymph nodes, however this was not excluded from our initial differential diagnosis and a solitary cystic squamous cell carcinoma in a cervical lymph node was suspected. Branchial cleft cyst and primary branchial cleft cyst carcinomas were also suspected because of their location in the same anatomical region.

#### **Surgical Procedure**

With the patient under oral endotracheal tube general anesthesia, a mid- horizontal incision overlying the cystic lesion was

made, upper and lower skin flaps with the platysma muscle raised, further clear dissection of the cystic wall was not possible, there was sever adhesions to sternomastoid muscle and muscles of the floor of the neck. Radical neck dissection was then carried out and the cystic mass removed together with other content of the neck. Gross inspection of the surgical specimen showed 5x4 cm in diameter cystic mass with thick cystic wall, the base of which had an indurated and thick fixed growth, when the cystic wall was incised, the inside surface of the growth showed thick granular base with friable surface and superficial ulceration (fig. 3).

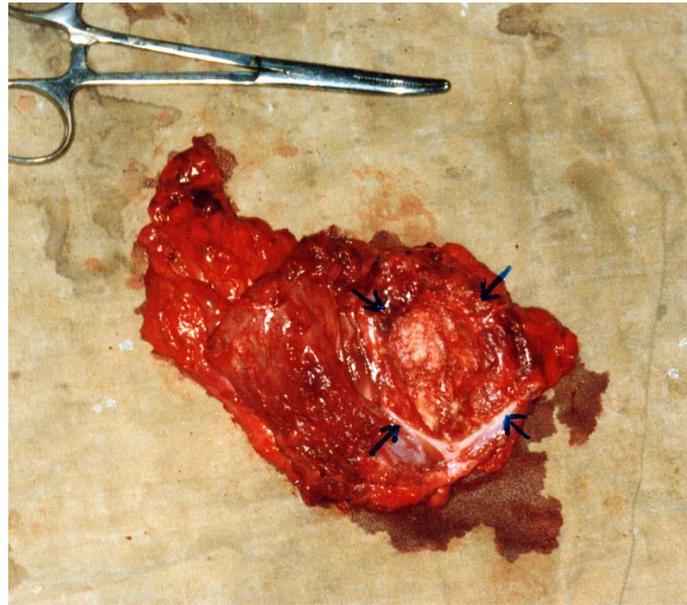
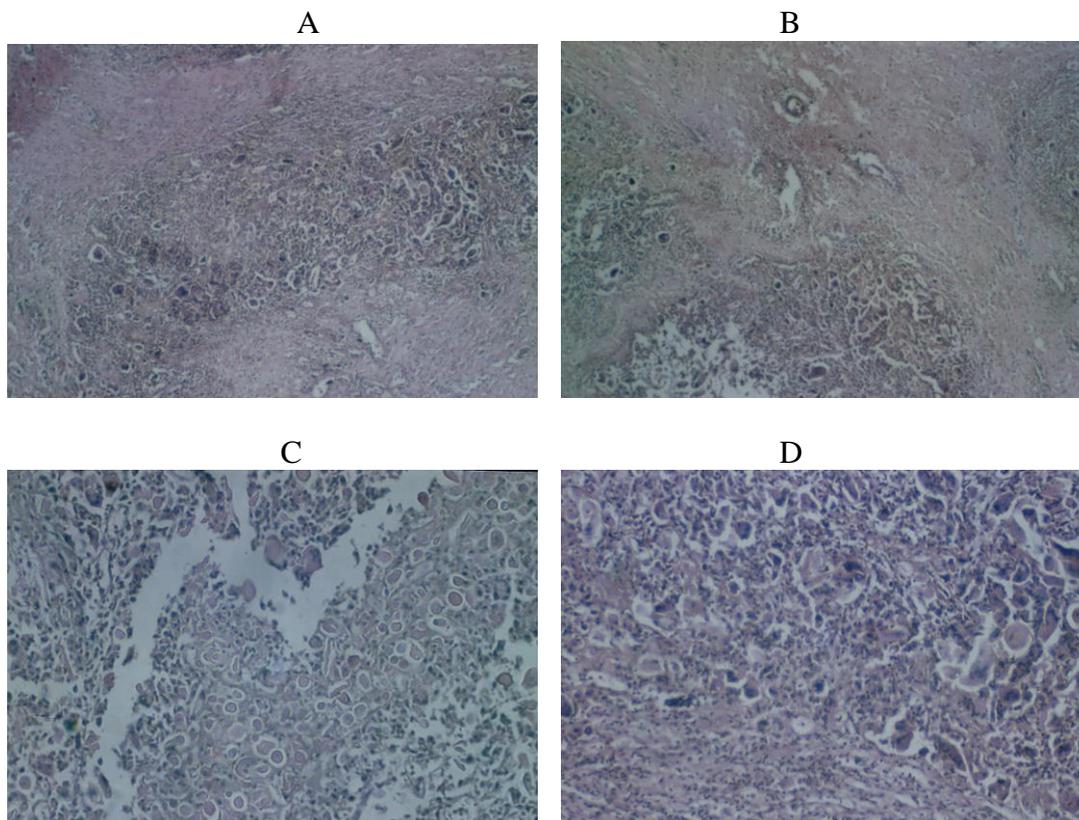


Fig. 3: Showing the gross surgical specimen. The boundary of the cystic mass is shown by the arrows, the base of the cyst is the granulomatous mass, the superficial surface of this mass is ulcerative, friable, shedding into the cystic cavity, this is shown by the white area in the center.

Histopathology of the specimen revealed mainly collagenous and skeletal muscle fibers showing multiple focal foreign body granulomas with large number of

foreign body giant cells, there was no cystic lining. No evidence of malignancy (fig. 4). Post-operative follow-up for twelve years showed no recurrence.



E

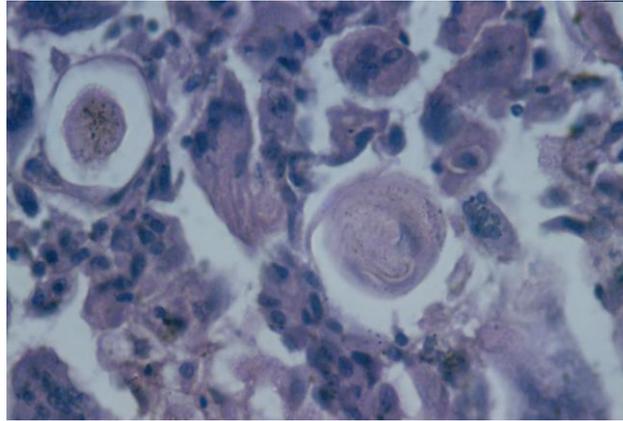


Fig. 4: A&B, Showing Collagenous and Skeletal Muscle fibers with Multiple focal foreign body granulomas. C&D, shows a large number of foreign body giant cells. Eosinophilic intracytoplasmic (keratin fragments ..?) Are seen within the giant cells, E.

## Discussion

Metastatic deposits in a cervical lymph node from oral squamous cell carcinoma will grow until the node is completely or partially replaced before the tumor passes to the next level of lymph nodes and or have an extra-capsular spread to surrounding soft tissue. However metastasis from squamous cell carcinoma often leads to various secondary changes in a lymph node such as necrosis, abscess formation, cystic and granulomatous changes<sup>4,6</sup>.

The Granulomatous reaction occurring simultaneously with metastatic squamous cell carcinoma in cervical lymph nodes draining laryngeal carcinoma has been described by Ophir et al<sup>5</sup>, upon finding of epithelioid granuloma in cervical lymph node, a malignant tumor of head and neck should be considered differential diagnosis possibility. However there was no mention if this represents a degenerative or regressive state in metastatic lymph node.

Westra et al<sup>7</sup> have pointed out that metastatic squamous cell carcinoma undergo a state of regression to form squamous cell granuloma after organ preservation therapy with radiotherapy and or chemotherapy. In a total of eight cases he showed foreign body giant cell reaction to keratin in the absence of viable tumor. Likewise, the data presented by Homa et al<sup>2</sup>, suggests that

radiation injury in squamous cell carcinoma may be manifested by marked hyperkeratinization of tumor cells, keratin compounds liberated induce sever foreign body giant cell reaction. However occasional foreign body giant cells are not infrequently observed in fields of degenerating or seemingly healthy non-irradiated squamous cell carcinomas<sup>2</sup>. Jolles and Koller<sup>3</sup> considered that hyperkeratinization of tumor cords occurring in squamous cell carcinoma is a degenerative process and not as a form of irradiation induced differentiation. This spontaneous regression of squamous cell carcinoma may also be indirectly supported by Kerabi<sup>4</sup>, who evaluated the efficacy of fine needle aspiration cytology in diagnosis of metastatic squamous cell carcinoma in head and neck, he stated that in four cases, granulomatous lymphadenitis was misdiagnosed from the smears which had necrosis and granuloma formation, however surgical biopsy of the nodes showed necrosis of malignant cells and foreign body granuloma formation in response to keratinous material. This FNAC was part of a diagnostic work-up and therefore the presence of keratin formation with subsequent foreign body granulomatous reaction has been induced in the absence of radiation. Spontaneous regression of metastatic

tumors in Xeroderma Pigmentosum has been mentioned by Henry et al<sup>1</sup> but this was with malignant melanoma and not in squamous cell carcinoma.

Solitary cystic squamous cell carcinoma in cervical lymph node is now regarded as a typical presentation of metastatic squamous cell carcinoma arising in the oro/nasopharynx. At the same time, the idea of a primary branchiogenic carcinoma has become a vanishing concept. Regauer, et al<sup>6</sup> demonstrated a distinct subset of metastatic nodes present as cystic masses, with most of the volume made up of a liquid center surrounded by a thin solid rim. This clearly demonstrate the possibility of a large cystic change in a metastatic lymph node that reaches the size of branchial cyst of the neck but keeping the histopathological appearance of squamous cell carcinoma at the periphery.

The experience with this family of Xeroderma Pigmentosum had shown that the primary squamous cell carcinoma of the face and oral cavity was slowly growing, non invasive and non-metastasizing. When the tumor was completely excised there was no

recurrence at the primary site. Histopathological examination of all the squamous cell carcinoma that had been excised from this patient were of the well-differentiated type.

Although in our presenting case, did not show, histologically, any remnant tumor tissue, we believe that, because of the above nature of the tumor, metastasis had gone a state of spontaneous regression and degeneration with the resultant of marked hyperkeratinization. Keratin formation has induced sever foreign body giant cell reaction, granuloma formation, with subsequent cystic change. This was clearly shown by the gross surgical specimen, which consisted of an extensive soft tissue mass infiltrating adjacent and underlying musculature, the superficial surface of this mass is yellowish white, granular and very friable and partly shedding to the cystic cavity, greatly resembling a large ulcerative oral squamous cell carcinoma (Fig. 3).

### Conclusion

Cervical foreign body giant cell granuloma with cystic change in Xeroderma Pigmentosum is reported for the first time.

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