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RESEARCH ARTICLE

AGGRESSIVE EPITHELIOID HEMANGIOENDOTHELIOMA OF THE THYROID: CASE REPORT

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Abstract

Epithelioid hemangioendothelioma (EHE) of the thyroid is an extremely rare disease; only five cases have been reported in the English literature to our knowledge. We report a case of a 57 year-old male presented with hematoma and recurrent thyroid mass after total thyroidectomy. Surgical exploration revealed the presence of a hematoma with tumor recurrence at the thyroid lodges. The histopathological and immunohistochemical findings indicated an EHE of the thyroid. With the rarity of Epithelioid hemangioendotheliom of the thyroid there is no standard therapy and the prognostic factors are unclear. Curative resection may be an effective treatment, further cases and studies are needed.

Introduction:-

Epithelioid haemangioendothelioma (EHE) was first described by Dail and Leibow in 1975 in lung and called « intravascular bronchioloalveolar Tumor ». After, in 1982 Enzinger and Weiss described it as a low-grade but potentially aggressive vascular neoplasm in soft tissu (1, 2). It occurs mainly in bone, soft tissue, lung and liver (3).
We report a case of primary malignant EHE of the thyroid very aggressive having recurred after surgical treatment with an unfavorable prognosis.

Case presentation

A 57-year-old non alpine mal was referred to our center for a recurrent post thyroidectomy hematoma. The patient had undergone a total thyroidectomy two months earlier that was complicated with a hematoma after nine days. Then he underwent two interventions for the drainage of the hematoma without success.

At admission the patient was pale and had anterior cervical mass (Figure 1). Nasofibroscopy showed a paralysis of the left vocal cord. The blood work revealed profound hypothyroidism with a moderate anemia. A full body Computed Tomography scan showed a 12 cm cervico-thoracic hematoma occupying the thyroid lodge (Figure 2), there was no other lesion elsewhere. Surgical exploration revealed the presence of a hematoma with tumor recurrence at the thyroid lodges.

The histological study found haemorrhagic zones and other hypercellular areas with the presence of abundant vascular structures of the capillary type. They show fibroblastic fusiform cells and mononuclear cells of histiocytoid appearance. Cellular pleomorphism and mitotic activity are moderate (Figure 3). The immunohistochemical study

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found a diffuse positivity of the tumor cells to anti-CK and CD34 antibodies (Figure 4). We present the other markers in Table 1. The diagnosis of hemangioendothelium epithelioid was retained.

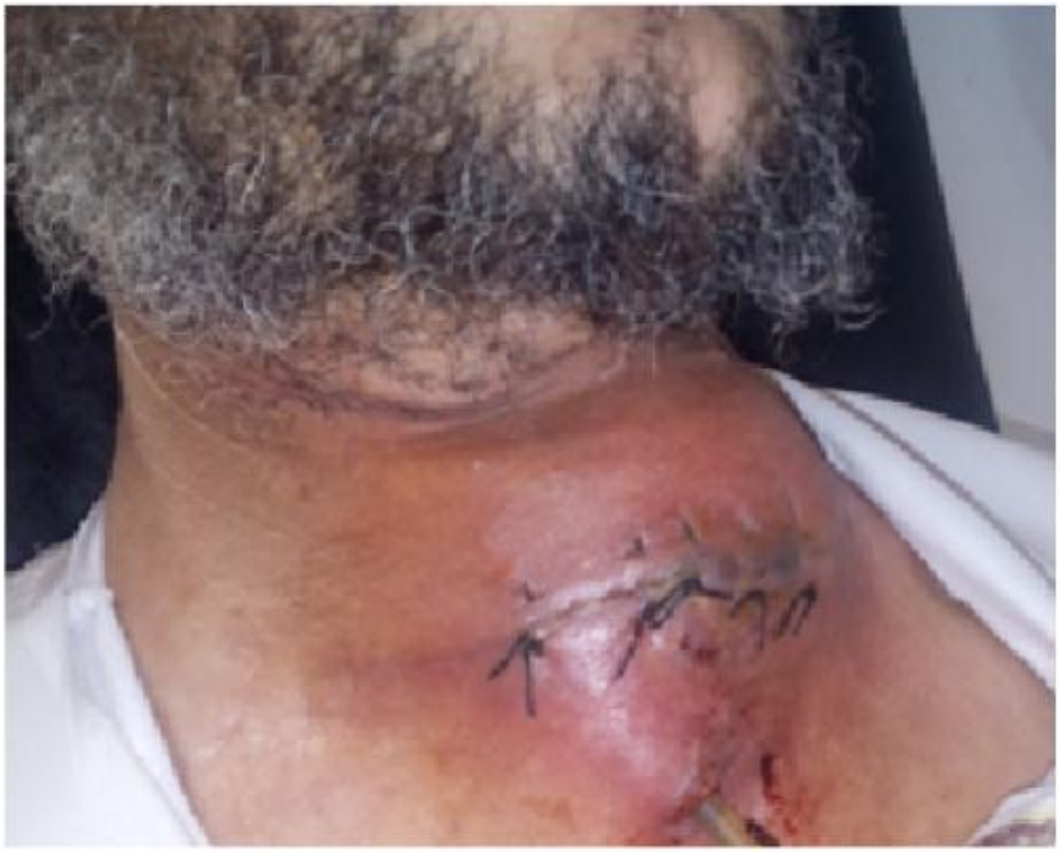

Figure 1:- Anterior cervical massrecurrent post thyroidectomy hematoma.

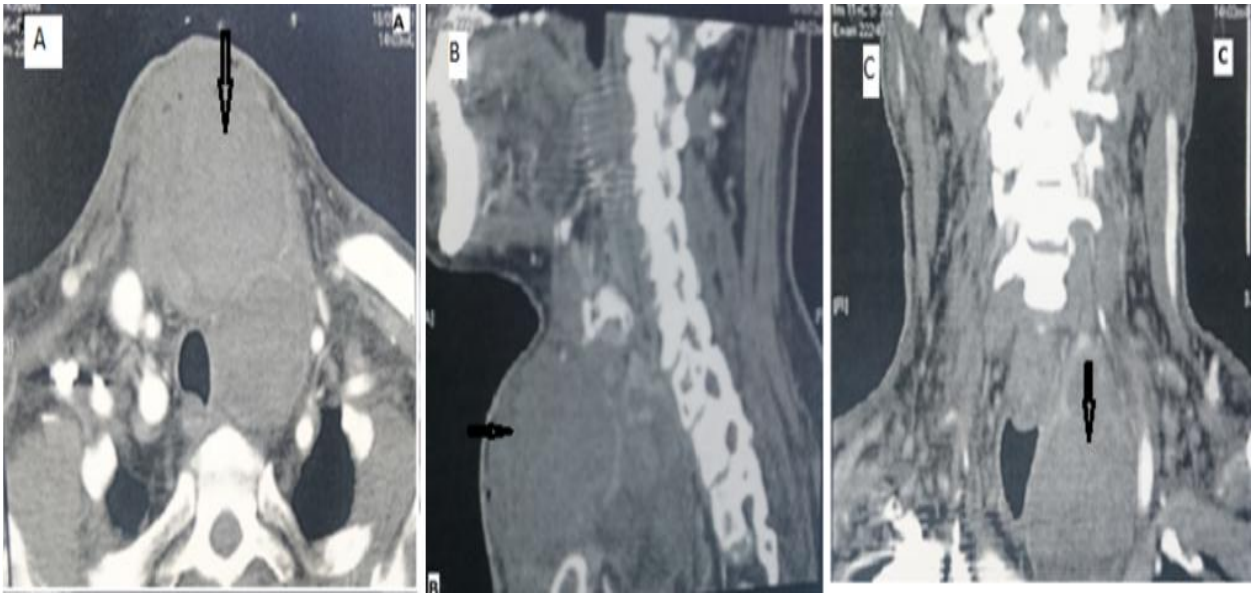

Figure 2:- Computed tomography of cervico-thoracic hematoma occupying the thyroid compartment. A: axial section. B: sagittal section. C: coronal section.

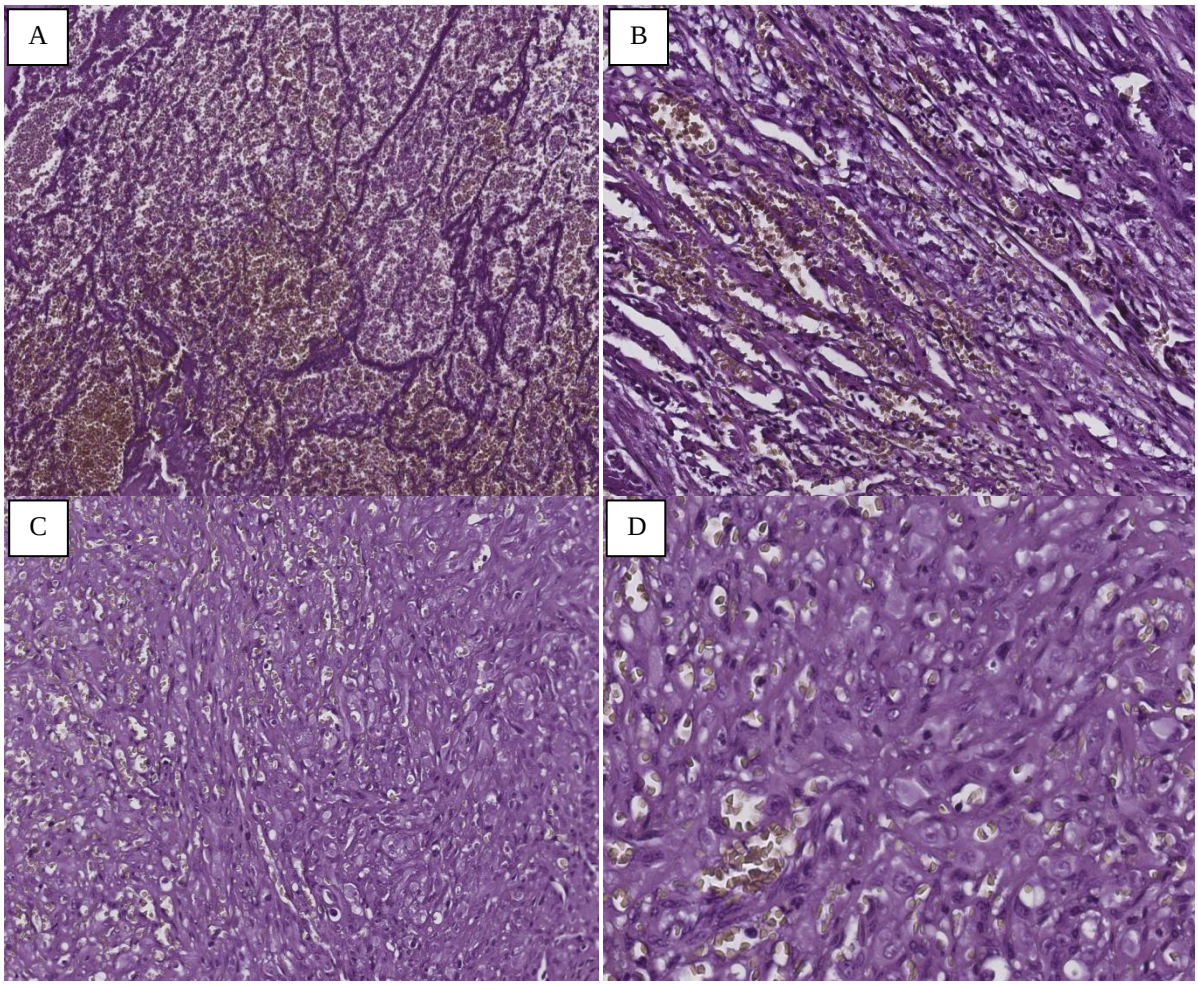

Figure 3:- Microscopic examination. A. Haemorrhagic zones (Hematoxylin and eosin stainx10). B. Vascular structures of the capillary type (X20). C. Cellular areas with the presence of abundant vascular structures of the capillary type (x20). D. Fibroblastic fusiform cells and mononuclear cells of histiocytoid appearance. Cellular pleomorphism and mitotic activity are moderate (x40).

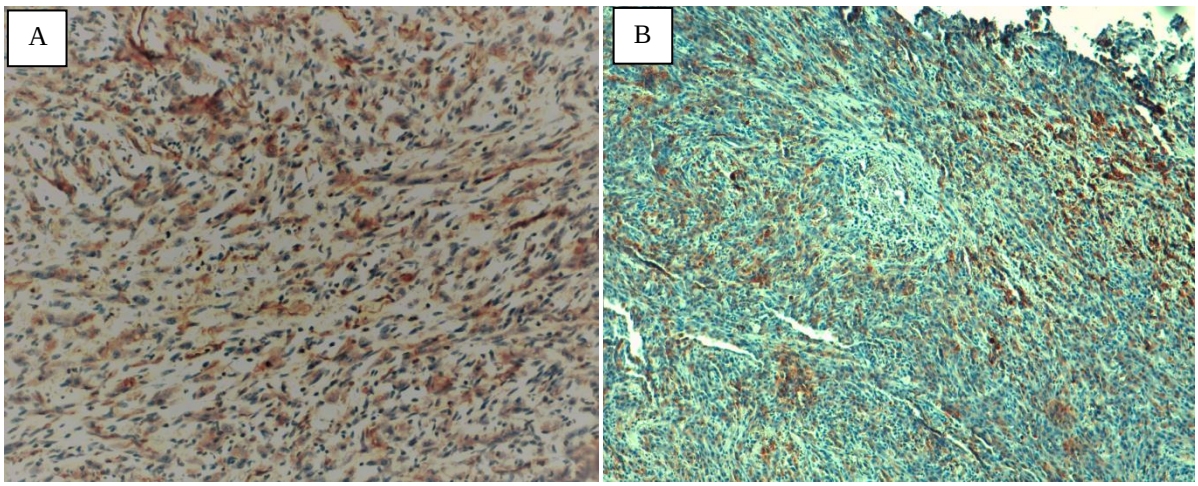

Figure 4:- Immunohistochemical study. A. Diffuse positivity of the tumor cells to the CD34 antibody (x10). B. Diffuse positivity of the tumor cells to the CK antibody (x10).

Antibody	Source	Clone	Tumor cell results
CD 34	Dako	QBEndo10	Positive
CD31	Dako	JC70A	Positive
EMA	Dako	E29	Focal positivity
FLI 1	Bio-SB	G146-12	Focal positivity
Vimentin	Dako	V9	Focal positivity
Cytokeratin	Dako	AE1/AE3	Positive
P53	Dako	DO-7	Focal positivity
Ki67	Dako	MIB-1	15%
Thyroglobin	Dako	Tg6	Negative
TTF1	Dako	8G7G3	Negative

Table 1:- Immunohistochemical reagents and staining profile of the tumor.

Discussion:-

EHE is a rare malignant angiocentric vascular neoplasm first described in 1982 by Weiss and Enzinger (4). It's composed of cords of epithelioid endothelial cells in a myxo-hyaline stroma with a presence of WWTR1-CAMTA1 fusion (4). The term EHE was initially proposed to designate a boderline heterogeneous group of vascular tumors with clinical, histological and evolutionary features that are intermediate between hemangiomas and angiosarcomas (3). EHE changed from an intermediate to malignant tumor in 2002 classification of World Health Organization (5). It has been described in bone, soft tissues and other organs such as lung, liver, spleen, brain, breast, heart, skin, peritoneum, and mediastinum (6, 3) but we found just five cases reported to date in the English literature in thyroid(1, 2,5, 7) (Table 2).

Author/ year	Age (year)	Sex	Thyroid tumor		Surgical procedure		Metas-tasis	Adjuvant therapy	Recur-rence	Out-come (follow-up period
			Loca-tion	**Size (mm)**	**Thyroid**	**Neck dissec-tion**				
Fujiwar/1998	56	M	Left lobe	30	Lobectomy	Left lateral	Lymph node	Radiation	(-)	Alive (11 months)
Siddiqui/1998	44	F	Right lobe	37	Lobectomy	Un-known	Un-known	(-)	(-)	Alive (24 months)
Hassan/ 2005	73	F	Right lobe	80	Thyroidec-tomy	Un-known	Un-known	INF-alfa	(+) 9 months	Dead (13 months
Shah/ 2016	35	F	Left lobe	27	Thyroidec-tomy	Left lateral	Lymph node	Plans for radiation and chemo-therapy	(-)	Alive (4 months)
Mayu/ 2017	74	F	Right lobe	21	lobectomy	Central	(-)	(-)	(-)	Alive (36 months)
Our case/ 2019	57	M	Un-known	Un-known	Thyroidec-tomy	(-)	(-)	Plans for radiation and chemo-therapy	(+) 2 months	Dead (2 months

Table 2:- Cases literature review of thyroid epithelioid hemangioendothelioma (EHE).

Thyroid localization of the EHE is extremely rare (2, 7), there are three cases in the English literature (1, 2). And two one case reported in the Japanese literature (5, 7). Most patients with thyroid EHE are women (male to female ratio, 2:4). Moreover, the literature reports the same observation for EHE of other organs, in which male to female

ratio is 1/4(9). In all cases, age is between 35 and 74 years with no predominance of any range. Unfortunately we do not have the patient's medical record before thyroidectomy, so we do not know the number, the size or the location of the nodules, but in the previous cases, tumor size was between 2.1cm and 8 cm, and all patients had a single nodule of the thyroid, contrary to other organs associated with multiple organ involvement in 36% of cases (8). The previous cases and our present case are summarized in Table 2.

Histologically, the tumor is consisting of chain and cords of epithelioid endothelial cells distributed in a myxohyaline stroma. The tumor cells centrifugally extend from the lumen of a large vessel to the surrounding tissue. This lumen contains tumor cells, necrotic debris and dense collagen (3). The cells have eosinophilic cytoplasm containing vacuoles called blister-cells and that deforms them and contains fragmented erythrocytes. Most of the time, the cells are low grade. This tumor express vascular markers especially CD34, CD31 and also FLI 1 and ERG transcription factor. In some cases we find a positivity of epithelial antigens CK7, CK8 and EMA (4).

At the molecular level the presence of CAMTA1 gene in 1p36 with WWTR1 gene fusion in 3q23-24 EHE was demonstrated (5, 4). This abnormality has been reported in the majority of EHEs at various anatomical sites, but he is not found in other epithelioid vascular tumors (5). It was confirmed in one case of thyroid EHE (1). In our case the search for molecular anomaly has not been realized.

The prognostic actors of EHE are the presence of pulmonary lesions, multiorgan involvement, age, and sex (5). In addition, mitotic activity (>3 mitotic figures/50 high-power fields) and size (>3.0 cm) have been used such as high risk factors in soft tissues (4). Although the thyroid EHE patient with the largest tumor (80 mm) died 13 months after the diagnosis , the other patients, whose tumor sizes were all <4 cm, had uneventful outcomes (5).In soft tissue 20-30% of tumors metastasize and about 15% of patients die (4). Our patient died within a few days of the anatomopathological diagnosis.

Abbreviation:-
EHE : Epithelioid haemangioendothelioma.
M: Male
F: Femal

Conflict of Interest:-
The authors declare that they have no competing interests.

Conclusion:-

With the rarity of Epithelioid hemangioendotheliom of the thyroid there is no standard therapy and the prognostic factors are unclear. Curative resection may be an effective treatment, further cases and studies are needed.

References:-

1. Shah AA, Ohori NP, Yip L, Coyne C, Antonescu CR, Seethala RR. Epithelioid hemangioendothelioma: a rare primary thyroid tumor with confirmation of WWTR1 and CAMTA1 rearrangements. Endocr Pathol. 2016;27:147–52.
2. Siddiqui MT, Evans HL, Ro JY, Ayala AG. Epithelioid haemangioendothelioma of the thyroid gland: a case report and review of literature. Histopathology.1998;32:473–6.
3. Bardouni A, Elouakili I, Ouchrif Y, Ouakrim R, Kharmaz M, Lamrani O, et al. Epithelioid hemangioendothelioma of the thigh: about a case. Pan African Medical Journal. 2014; 18:106.
4. 4-Fletcher CD, Hogendoorn P, Mertens F, Bridge J. WHO Classification of tumours of soft tissue and bone. 4th ed. Lyon: IARC Press; 2013.
5. Ono M, Kasuga Y, Uehara T, Oda Y. Epithelioid hemangioendothelioma of the thyroid: a case report. Surgical Case Reports. 2017 ; 3:18.
6. Hassan I, Barth P, Celik I, Hoffmann S, Langer P, Ramaswamy A, et al. An authentic malignant epithelioid hemangioendothelioma of the thyroid: a case report and review of the literature. Thyroid. 2005;15:1377–81.
7. Fujiwara K, Miura M. Epithelioid hemangioendothelioma of the thyroid gland: a case report. Pract Otorhinolaryngol. 1998;91:195–8.

8. Lau K, Massad M, Pollak C, Rubin C, Yeh J, Wang J, et al. Clinical patterns and outcome in epithelioid hemangioendothelioma with or without pulmonary involvement: insights from an Internet registry in the study of a rare cancer. Chest. 2011;140:1312–8.
9. Sardaro A, Bardoscia L, Petruzzelli MF, Portaluri M. Epithelioid hemangioendothelioma: an overview and update on a rare vascular tumor. Oncol Rev. 2014;8:259.