



## LETTER TO THE EDITOR

## Composite hemangioendothelioma on the neck

To date, only 27 cases of composite hemangioendothelioma (CHE) have been reported. To further broaden the clinicopathological spectrum of CHE, we present the first report of a case of CHE in the right neck region.

A 46-year-old woman presented with an isolated asymptomatic mass on the right side of her neck; she had first noticed the mass 4 years previously. Physical examination showed an oval neoplasm. The results of a blood film analysis, kidney and liver function tests, urine analysis, and chest radiography were normal. Cervical magnetic resonance imaging showed a mass with a diameter of 4.8 cm in the subcutaneous tissue (Fig. 1A). This mass was surgically excised with a 1 cm safety margin.

Gross examinations showed an oval, firm, grayish-red-colored mass with dimensions of 4.8 cm × 3.7 cm × 2.1 cm. The cut surface was whitish-gray. Microscopic examination showed a lesion composed of vascular spaces and solid areas, with a complex mixture of different histological patterns and poorly defined margins. The most striking feature on low-power examination was the variability of appearance across different areas. The predominant histological components were epithelioid hemangioendothelioma (approximately 60%), retiform hemangioendothelioma (approximately 30%), and papillary intralymphatic angioendothelioma (approximately 10%) (Fig. 1B).

The epithelioid hemangioendothelioma components showed nests of round to slightly spindle-shaped epithelioid cells surrounded by myxohyaline stroma. Many cells showed intracytoplasmic vacuoles, and some of these vacuoles contained erythrocytes. In one small area, the vacuolization was so intense that the cells seemed to have adopted a lipoblastic appearance.

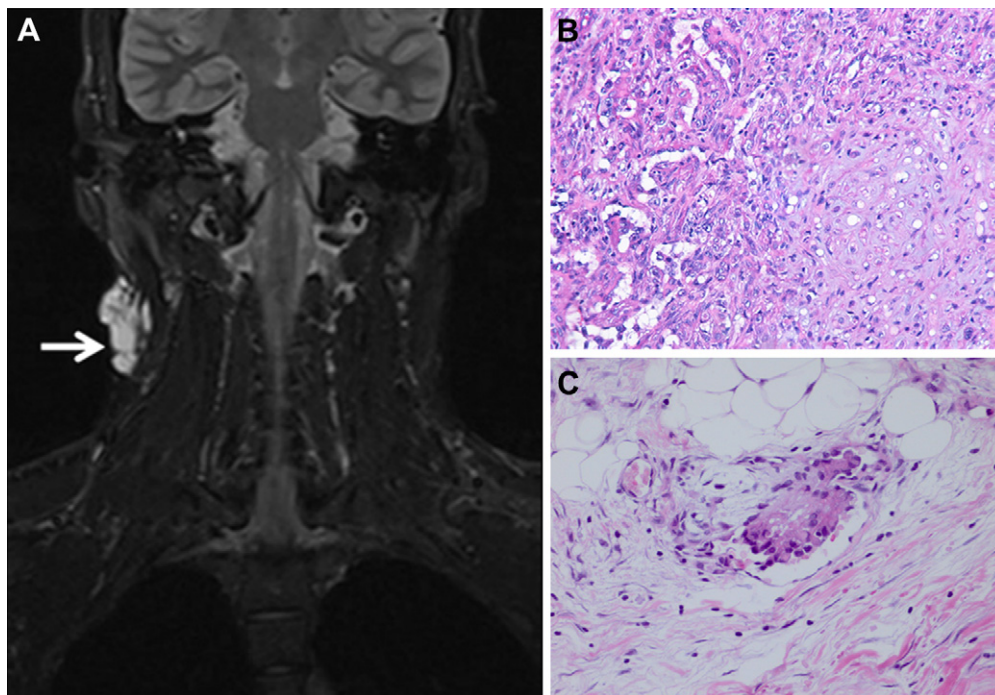
Retiform hemangioendothelioma components were characterized by elongated and narrow arborizing

vascular channels with a striking resemblance to the normal rete testis, and lined with plump and hobnailed epithelioid cells. Papillary plugs of the lesions were composed of papillary tufts with hyaline cores lined by hobnail endothelial cells extending into the lumen, resembling a Dabska tumor (Fig. 1C).

The tumor showed no evident nuclear atypia or typical necrotic areas. Mitotic figures were rare. Immunohistochemical findings revealed that the neoplasm was positive for CD34, CD31, and vimentin (VIM), and negative for cytokeratin-7 and S-100. On the basis of the presence of these variable histological characteristics, a final diagnosis of CHE was made.

CHE is a rare type of hemangioendothelioma that was first described as a separate entity by Nayler et al. in 2000 [1]. The neoplasm is characterized by the simultaneous presence of different histological patterns, the percentages and distributions of which differ in each case. In the 27 previously reported cases, CHE mostly affected adults (18 cases, 72%) [1–5], but it has also been reported in children (7 cases, 28%) [1,2,5]. The incidence in female patients is higher than that in male patients, with a female:male ratio of approximately 1.7:1.

Most cases of CHE (20 cases, 74.1%) have shown a predilection for the extremities. The tumors were found in the tongue and the cheek mucosa in two cases (7.4%) [1], in the mandibular vestibule in one case (3.6%) [2], on the back in one case (3.6%) [3], in the mediastinum in one case (3.6%) [4], and in the hypopharynx in two cases (7.4%) [5]. The tumor in our case was located in the right neck region, a hitherto undescribed primary tumor site. We suggest that CHE may affect a wider range of body locations than previously reported.



**Figure 1.** (A) Magnetic resonance enhancement presented an oval area of enhanced signaling in the right neck. (B) This complex lesion had areas consistent with retiform haemangioendothelioma (left) as well as more solid areas consistent with epithelioid haemangioendothelioma (right). HE stain, 200 $\times$ . (C) Intravascular papillae with collagenous cores similar to those seen in Dabska's tumour. HE stain, 200 $\times$ .

## References

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