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Pituitary Lesion of Unknown Origin: Think Epithelioid Angiosarcoma

Léopoldine Bricaire,[1]* Chiara Villa,[4,8]* Mathilde Brière,[1] Myriam Edjlali,[5,6] Frédérique Larousserie,[2,5] Marguerite d'Ussel,[7] Jérôme Bertherat,[1,5,8] Pascaline Boudou-Rouquette,[3] and Lionel Groussin[1,5,8]

Departments of [1]Endocrinology, [2]Pathology, and [3]Oncology, Hôpital Cochin, Assistance Publique-Hôpitaux de Paris, 75014, Paris, France; [4]Department of Pathology, Hôpital Foch, 92151, Suresnes, France; [5]Université Paris Descartes, Sorbonne Paris Cité, 75005, Paris, France; [6]Department of Neuroradiology, INSERM UMR894, Hôpital Sainte-Anne, 75013, Paris, France; [7]Department of Emergency Unit, Hôpital Saint-Joseph, Paris, 75015, France; and [8]Institut Cochin, Inserm Unité 1016, Centre National de la Recherche Scientifique Unité Mixte de Recherche 8104, Descartes University, 75014, Paris, France

A 72-year-old woman presented in the emergency unit of Saint-Joseph Hospital, Paris, France, with subacute intracranial hypertension syndrome evolving over the last 2 weeks. At admission she was confused, and on physical examination she presented a left third nerve palsy. Laboratory analysis found hypernatremia at 150 mmol/L (reference range, 135 to 145 nmol/L) and panhypopituitarism confirmed by hormonal investigations. The patient had no relevant medical history. A whole-body clinical examination, particularly of the skin and mucous membranes, did not find any specific lesion. Brain magnetic resonance imaging revealed a 4-cm heterogeneous enhancing sellar and suprasellar lesion, with substantial mass effect on the chiasm and third ventricle (Fig. 1). A reactive brain edema was seen around the mass on the axial T2 and coronal fluid-attenuated inversion recovery sequences, which is not a typical feature of macroadenoma and could suggest, although rather nonspecific, an unusual lesion. Whole-body computed tomography scan and ^{18}F-fluorodeoxyglucose positron emission tomography/computed tomography scan excluded a secondary lesion from a cancer arising elsewhere in the body, suggesting a primary sellar or suprasellar lesion. The mass was transsphenoidally partially resected, and the patient received gamma-knife radiotherapy on the residual lesion. Unfortunately, the tumor progressed after radiotherapy, and the patient died 3 months later of intracranial hypertension syndrome.

Histologic examination revealed a hypercellular tumoral proliferation formed by large atypical monomorphic cells with large vesicular nuclei, prominent nucleoli, and abundant light eosinophilic cytoplasm. Tumor cells were arranged in solid sheets [Fig. 2(A)]. Numerous mitosis and necrosis were observed. No features of vascular differentiation, for example intracytoplasmic lumina or areas of conventional angiosarcoma, were observed.

For this reason, a broad immunohistochemical panel was applied searching for epithelial, neuroendocrine, melanocytic, lymphoid, mesenchymal, germ cell, and neuroglial differentiation. All markers were negative, with the exception of ERG [Fig. 2(B)], CD31 [Fig. 2(C)], cytokeratin AE1/AE3 (Golgi-like patchy expression) [Fig. 2(D)], and CD34. This immunophenotype excluded a carcinoma, a lymphoid, melanocytic, germ cell, or neuroglial proliferation, and the diagnosis of sellar and suprasellar epithelioid angiosarcoma was made.

*These authors contributed equally to this study.

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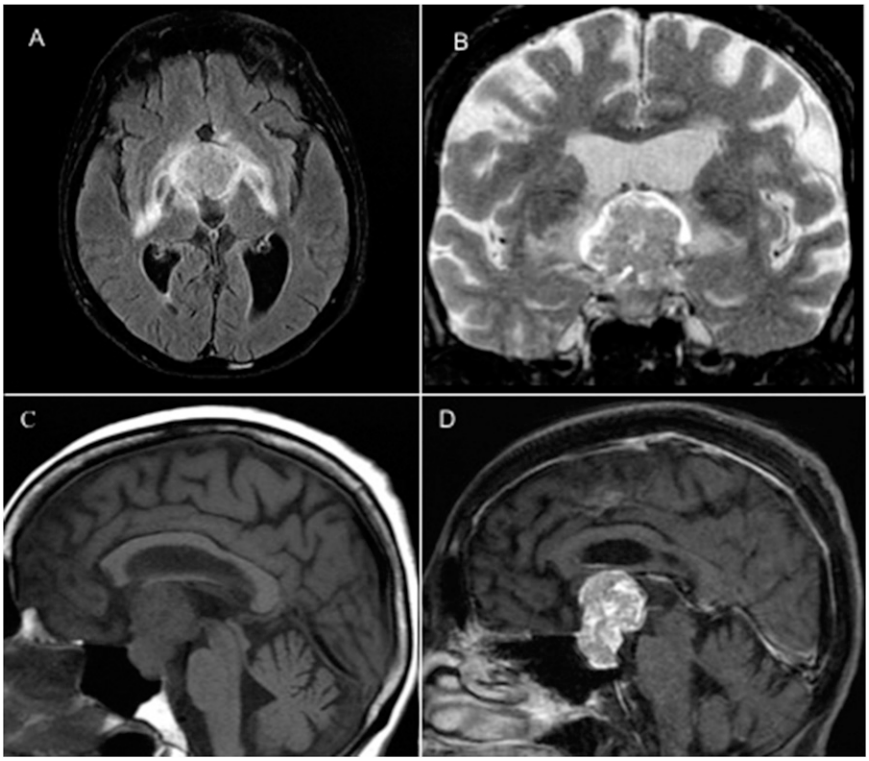

Figure 1. Brain magnetic resonance imaging findings. (A) Axial, (B) coronal, and (C and D) sagittal sections of (A) T2 fluid-attenuated inversion recovery, (B) T2, (C) T1 pregadolinium, and (D) postgadolinium, showing a 4-cm heterogeneous enhancing sellar and suprasellar lesion, with substantial mass effect to the chiasma and third ventricle.

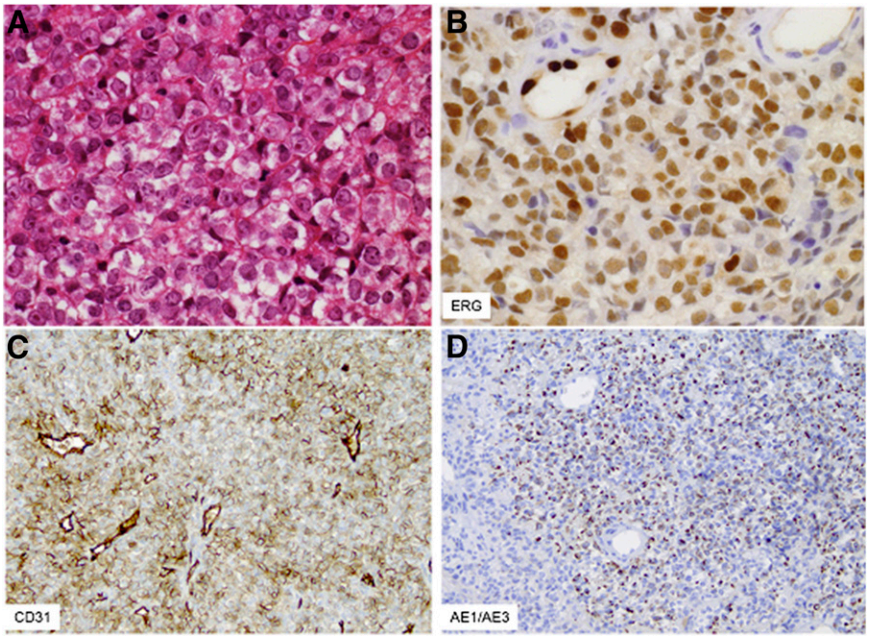

Figure 2. Pathologic features of epithelioid angiosarcoma. Magnification of ×400 with (A) hematoxylin and eosin stain, (B) ERG immunostaining, and (C) CD31 and ×200 magnification with (D) pan-keratin AE1/AE3 immunostainings. Malignant proliferation showing large monomorphic cells with some epithelioid (A1E/AE3) or angiocentric aspects without clear vascular differentiation. The degree of cellularity, atypia, and mitotic activity is high. ERG and CD31 immunoexpression prove endothelial differentiation.

In front of an aggressive sellar and suprasellar lesion with a subacute clinical presentation, the main diagnoses are a pituitary carcinoma or aggressive adenoma, a pituitary metastasis of a carcinoma of unknown origin, or a solid craniopharyngioma. This case shows that vascular markers should be done in front of an aggressive sellar lesion with unexpected histologic features. Vascular markers allowed us in this case to make the unusual diagnosis of primary epithelioid angiosarcoma. It has been rarely described in the central nervous system but never, to our knowledge, in the pituitary gland.

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Address all correspondence to: Léopoldine Bricaire, MD, Department of Endocrinology, Cochin Hospital, 27, rue du Faubourg Saint Jacques, 75014 Paris, France. E-mail: leopoldine.bricaire@aphp.fr.

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