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Malignant Transformation of Multiple Dermal Cylindromas

Cylindromas are benign tumors appearing as small solitary slow growing nodules on the head and neck. Multiple cylindromas may form a turban tumor. Here, we report an unusual case of multiple cylindroma with transformation to cylindrocarcinoma. The patient is a 61-year-old woman who developed a cylindrocarcinoma on a pre-existing cylindroma of head and neck, with deformity of head due to soft tissue masses, lytic lesions of scalp with invasion to brain, and destruction of orbit leading to unilateral visual loss.

Keywords: multiple cylindroma, malignant transformation, imaging

Introduction

Dermal cylindromas are benign tumors assumed to differentiate towards apocrine sweat glands of the skin. Most arise as small solitary slow growing nodules in the head and neck region. Multiple cylindromas may form a so-called turban tumor in the autosomal dominant Brooke-Spiegler syndrome associated with multiple tricho-epitheliomas.¹ There are only 31 cases of multiple transformation of dermal cylindromas in the literature.² We report a patient with malignant cylindrocarcinoma with invasion to the skull and brain, and unilateral visual loss.

Case report

A 61-year-old woman with a tumoral mass in the frontal and occipital parts of the scalp since 15 years ago presented with sudden growth of the mass over the last three years plus loss of vision in her left eye and grade II papilledema. Family history of the patient was negative for these kinds of tumors. In clinical examination, multiple tumoral lesions were seen in the right frontal, left occipital and left zygomatic bones extending to the left orbit and left side of the neck with red discoloration and varicose veins on the face and scalp, and head deformity (Figure 1). Two maculopapular lesions were seen on her left hand.

Since the patient did not consent, surgery was not performed. Skull x-ray showed large lytic lesions with bony nests and speculations in the affected parts (Figure 2). In carotid and vertebral angiography, a highly vascular mass with aneurysmal changes, vascular displacement, and AV shunts with tortuous and dilated vessels were seen in the affected parts too (Figure 3). A huge heterogeneous soft tissue mass in the right temporo-parieto-occipital and left fronto-temporo-parietal areas was seen with extensive bone destruction (of both the outer and inner tables) and bone expansion. Invasion of the brain parenchyma in the left occipital and posterior parietal lobes, right cerebellum, right CPA, left cerebellum, right temporal lobe, and left orbit with displacement of the globe inferiorly and medially in front of left maxillary sinus were also seen. Posterior cervical extension with invasion and destruction of the first cervical vertebra was seen too (Figures 4 and 5).

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Fig 1: Photograph of the patient shows multiple tumoral lesions in right frontal, left occipital, left frontal and left zygomatic bone with extension to left orbit and left side of neck and red discoloration of varicose veins on face and scalp and head deformity.

particles of 5×2.5×1.5 centimeters. Microscopic examination revealed a neoplasm with cuboid to round cells. These cells had clear homogenous cytoplasm with conglomeration which made a tubular appearance containing secretions.

Discussion

Cylindromas are benign tumors, and it is believed that they show a differentiation towards apocrine sweat glands. Histologically, nests of basaloid cells with partly clear and partly dark nuclei are surrounded by a hyaline sheath composed of basement membrane components such as collagen IV and laminin.³ The first description of multiple cylindromas of the head and abdomen dates back to 1842 by Ansell.⁴ In 1929, Wiedemann first described a malignant cylindrocarcinoma characterized by loss of the typical 'jigsaw pattern' and of the peripheral palisading of the basaloid cells, and polymorphous clear cells with prominent nucleoli.⁵

The skull biopsy contained multiple specimens of gray fragile and firm tissue to the best of our knowledge, there are only 31 cases of malignant cylindrocarcinoma described in the literature up to now.² Malignant transformation affects multiple cylindromas more often than solitary ones.⁶ The age of patients with cylindrocarcinoma in the published reports ranges from 50 to 96 years with a slight female predominance. None of the reported cases are as large as our case, with widespread extension to soft tissue, bony structures, brain parenchyma, and orbit (dislocating the globe), TM joint, cervical canal, and the cord. Reported cases were located on the skull with local infiltration and destructive growth.^{7,8}

Clinical signs of malignant transformation are ulceration, rapid growth, bleeding, and blue to pink discoloration of nodules.⁹ Metastatic spread to other organs has been reported in 11 patients, nine of whom died of metastatic disease. Lymph nodes, stomach, thyroid, liver, lung, and bones were affected in these metastases.^{4,5,10-16}

In conclusion, malignant transformation of dermal cylindroma is rare. It should be however, considered in clinically altered tumors, especially those arising from multiple cylindromas of the head or trunk. A close follow-up should be performed because metastatic spread is frequent. Radiotherapy seems to be an option in inoperable tumors.

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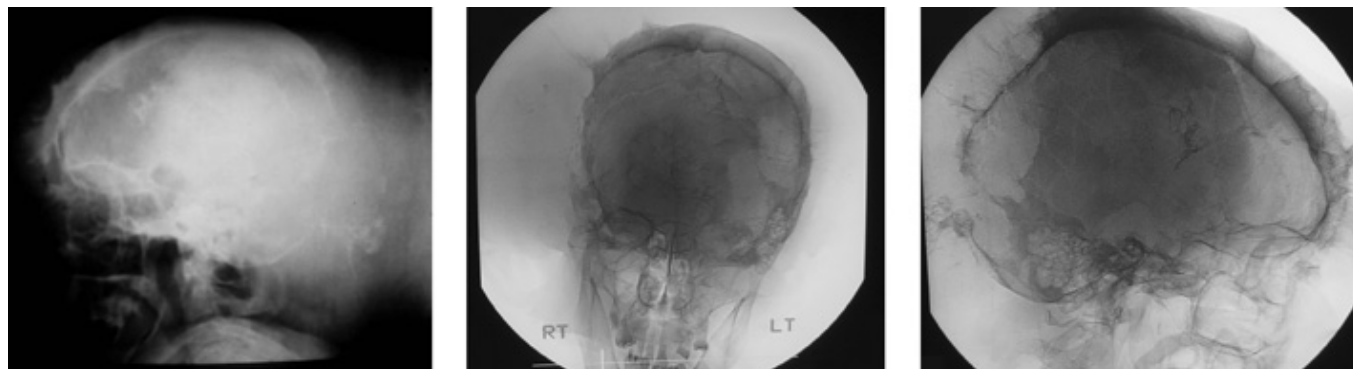


Fig 2: Skull AP and LAT x-ray shows a large lytic lesion with bony nests and speculation in affected parts of skull including right frontoparietal and left occipital bones, left zygoma and orbit with a huge soft tissue mass overlying the bony changes especially in the occiput.

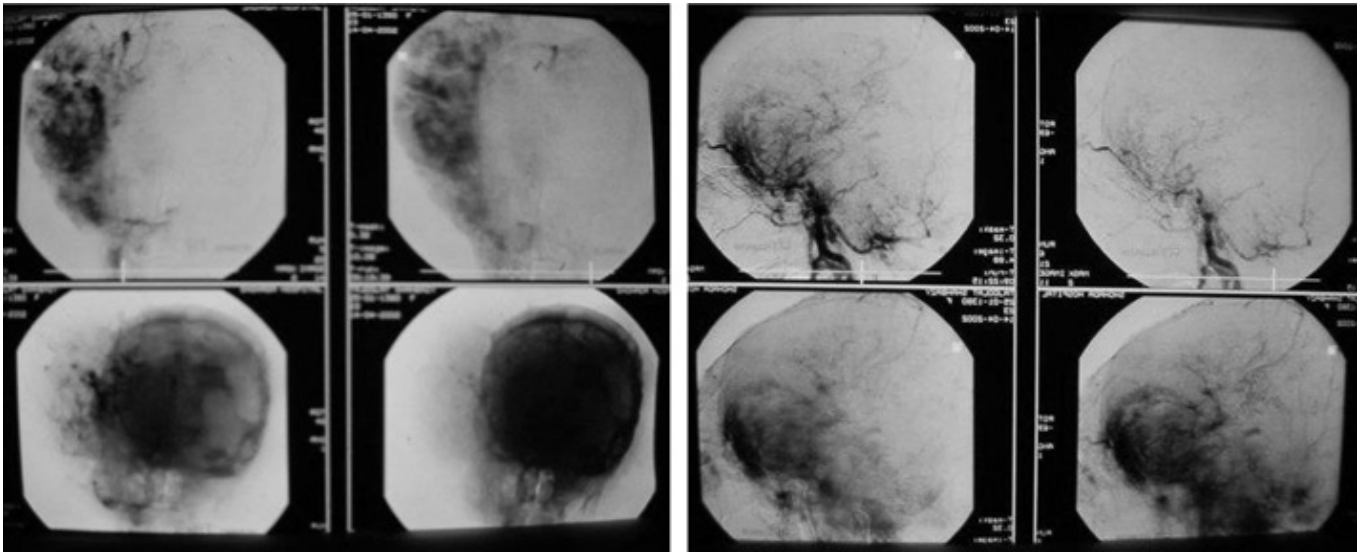


Fig 3: DSA angiography from right ICA shows a highly vascular mass with aneurismal changes, vascular displacement, AV shunts with tortuous and dilated vessels in affected parts.

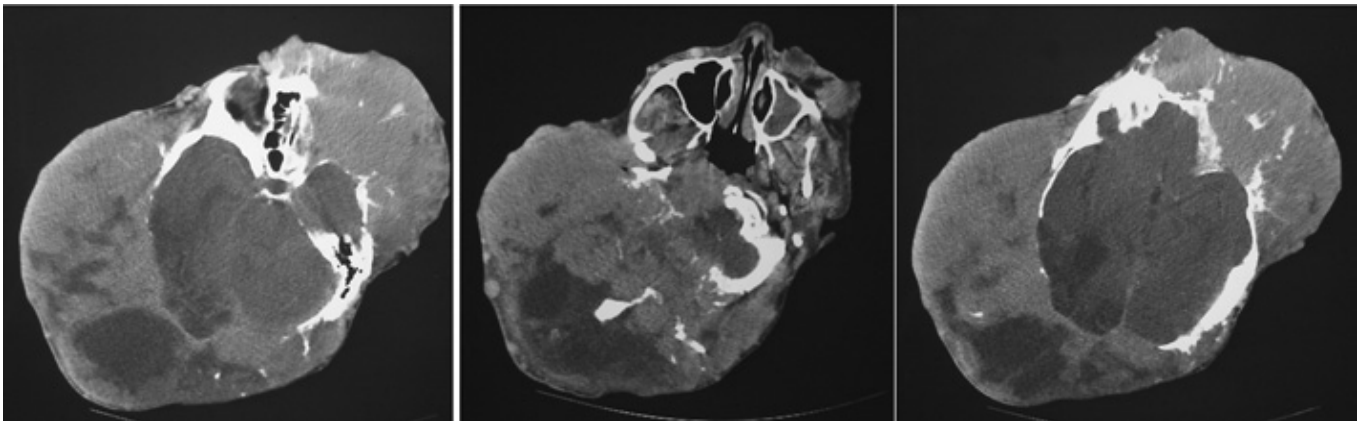


Fig 4: Axial brain CT scan shows a huge heterogenous soft tissue mass in right temporoparietooccipital and left frontotemporoparietal areas of the skull with extensive bony destruction in both outer and inner tables with expansion of bone, invasion to brain parenchyma at left occipital and posterior parietal, right cerebellum, right CPA, left cerebellum, right temporal lobe, left orbit with displacement of globe to inferior and medial in front of left maxillary sinus. Also extension to posterior of cervical canal with invasion and destruction of first cervical vertebra.

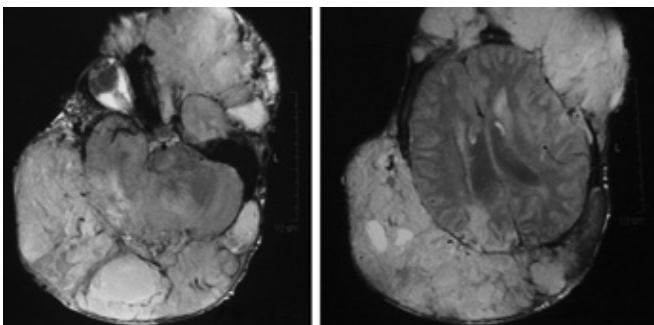


Fig 5: Axial proton density brain MRI shows extensive hypersignal soft tissue mass with involvement of brain parenchyma in right occipital lobe.

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