Nevus planus of the skin associated to jugulo-tympanic paraganglioma. A new neurocutaneous syndrome?

SIR,—The metamic association of a cutaneous planus angioma and a spinal angioma under the name of Cobb’s syndrome is well known. However, the association of cutaneous angioma with very vascular tumours, such as haemangioblastoma of the spinal cord, has never been described.

For several years, we have followed a female patient who has a cervical cutaneous nevus planus associated with an ipsilateral jugulo-tympanic paraganglioma, a tumour that is highly vascularised. This patient was first seen when she was 60 years old. For 15 years there had been very slow progressive hoarseness, dysphagia, left deafness and occasional left hemifacial spasm. When she was 46 years old, she had been seen by an otologist who saw a very vascularised polypus of the middle ear and told her that it was the cause of her deafness. She had, in addition to the cutaneous angioma (figure), hypaesthesia of the first branch of the fifth cranial nerve and a complete lesion of the 8th, 9th, 10th, 11th, and 12th cranial nerves, all on the left side. Carotid and vertebral angiography and CT-scan showed a tumour that was filling the left middle ear, destroying the petrous pyramid and foramen jugular and extending towards the posterior fossa from the free edge of the tentorium to the foramen magnum. The radiological findings were consistent with the tumour being a glomus tumour. Considering the age of the patient, the nature and extent of the lesion, the very slow progression of symptoms and the almost normal functional state despite the wide involvement of the left low cranial nerves, we considered that surgical treatment was not advisable. Three years after the diagnosis, the patient is in good health and her neurological symptoms and signs have not changed. Although we do not have pathological evidence of the nature of intracranial tumour, the radiological findings and the ear vascular polypus are characteristic of a glomus tumour.

We think that this is the first time that a cutaneous angioma associated with a jugulo-tympanic paraganglioma has been reported. This association may constitute another neurocutaneous syndrome, and also raise the possibility that a neuroectodermal dysplasia may be the primitive substratum of jugulo-tympanic paraganglioma.

References

Figure. Photograph of the patient showing the cutaneous angioma. As can be seen, there is atrophy of the left half of the tongue due to hypoglosal palsy.

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