

pyogenic granuloma, basal cell carcinoma, and inflammatory conditions, as these would present other types of vessels.

To date, 3 reports have been published on the dermoscopic features of skin metastases from internal malignant tumors. De Giorgi et al.⁴ and Oiso et al.⁵ described atypical polymorphous vascular patterns in skin metastases from a recurrent thyroid carcinoma and in a skin metastasis from a possible occult breast cancer. But it was Mun et al.⁶ who first described the dermoscopic features of SMJN that had arisen from an adenocarcinoma of the pancreas. Recently, Chernoff et al.⁷ published an article in which they analyzed the vascular patterns of skin metastases; the most typical dermoscopic findings were serpentine vessels, linear irregular vessels, and unstructured homogeneous areas of pink color.

The dermoscopic vascular pattern is considered to be a pathognomic phenomenon of neovascularization related to neoplastic growth.⁴ In the present case, dermoscopic examination of the umbilical lesion revealed a polymorphous vascular pattern that raised our suspicion of malignancy and led us to perform a skin biopsy that identified the presence of metastatic adenocarcinoma.

When an umbilical nodule is observed, a metastatic malignant tumor must be excluded as this may be its first clinical manifestation and indicates a poor prognosis. As far as we are aware, this is the second case report of the dermoscopic features of SMJN. The aim of this report is to increase the suspicion of a metastatic tumor when dermoscopic examination of an umbilical lesion reveals a polymorphous vascular pattern. Dermoscopy is now an essential additional tool for dermatologic diagnosis.

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Schwannoma of the Tongue[☆]



Schwannoma de la lengua

To the Editor:

The schwannoma, neurilemmoma, or neurinoma is a benign tumor that arises from the Schwann cells of the cranial and peripheral nerves. It appears in individuals between 20 and 50 years of age and is equally common in men and women.¹

Schwannomas are usually sporadic, but in some cases they are associated with neurofibromatosis type II, schwannomatosis, Carney complex, or, more rarely, with radiation therapy, with a latency of up to 50 years (acoustic schwannomas).²

Around 25% of extracranial schwannomas arise on the head and neck, and approximately 1% are intraoral.^{3,4}

Our patient was an otherwise healthy man of 52 years of age, with no past medical history of interest. He presented a single tumor that had appeared 18 months earlier on the dorsum of tongue. The tumor was occasionally painful. There

was no history of surgery or trauma to the area. Physical examination revealed a nodule of 2 mm diameter on the dorsum of the tongue. The lesion was slightly erythematous, shiny, well-defined, slightly tender, and had a smooth surface and a rubbery consistency (Fig. 1).

Simple excision of the lesion was performed and histopathology revealed a clearly circumscribed tumor composed of fascicles of spindle-shaped cells, lying beneath the mucosa of the tongue (Fig. 2). At higher magnification, elongated cells were visible; the cells did not show atypia and their nuclei were arranged in line forming the typical "Verocay bodies" (Fig. 3). Immunohistochemistry was positive for S100. A diagnosis of classic schwannoma was made based on these findings.

After excision of the lesion, the wound was closed by direct suture and there have been no surgical complications or recurrence after 6 months of follow-up.

Schwannoma is rare on the tongue. Cohen et al.¹ performed a review of schwannomas on the tongue published in journals indexed in PubMed and found 126 cases over a period of 56 years (including 2 cases published in the same article).

Schwannoma presents clinically as a firm, slow-growing tumor that can reach a variable size. Symptoms depend on the site and size, and the tumor can be painful,³ as in the case we have presented, or asymptomatic. When situated on

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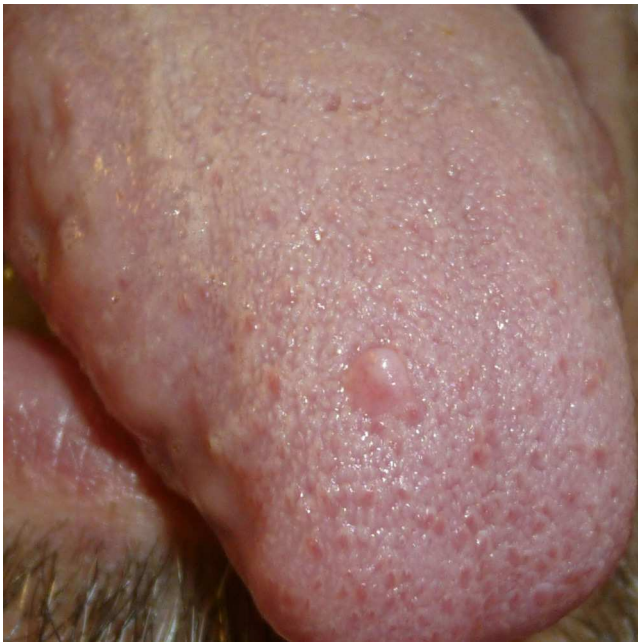


Figure 1 Well-defined, erythematous nodule of 2 mm diameter on the dorsum of the tongue.

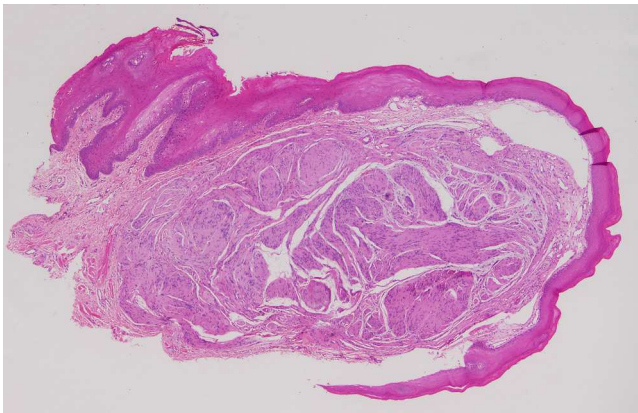


Figure 2 Histopathology revealed a clearly-circumscribed submucosal tumor (hematoxylin and eosin, original magnification $\times 2$).

the tongue, some patients describe inflammation, fasciculations, or weight loss associated with difficulty swallowing. Very rarely the lesion can grow rapidly and ulcerate.⁵

The definitive diagnosis is histological, by observation of a well-defined, encapsulated tumor formed of spindle-shaped cells that can be arranged in 2 patterns. The "Antoni A areas", which are more cellular and contain the characteristic Verocay bodies, formed of elongated cells with no atypia; the cell nuclei are arranged in line and their cytoplasm is fused into eosinophilic masses. The "Antoni B areas" contain randomly distributed cells in a myxomatous stroma.¹⁻⁶ Immunohistochemistry is positive for S100. Depending on the morphological variant, schwannomas can be classified into classic, cellular, plexiform, ancient (degenerated), and melanotic.²

Loss of function of the merlin protein (coded by gene *NF2*)³ appears to be the cause of the appearance of

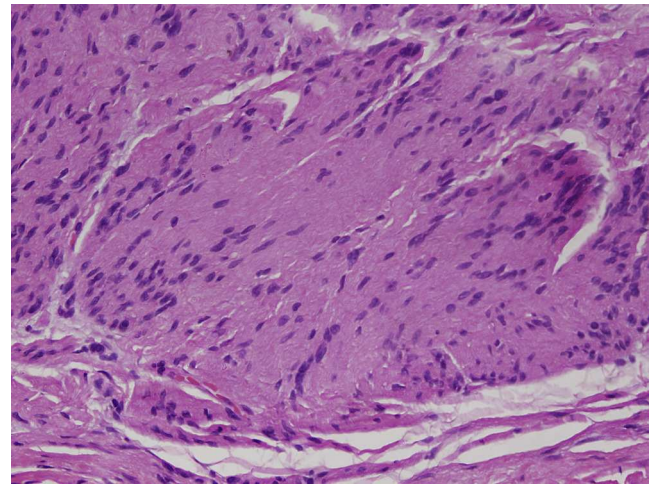


Figure 3 Cells grouped in palisade, forming the structures called Verocay bodies (hematoxylin and eosin, original magnification $\times 20$).

schwannomas. This protein has complex functions that are essential to various pathways in the nucleus and in the cell membranes. Loss of merlin function in the nucleus leads to an increased expression of membrane proteins, including integrins and growth factor receptors. These growth factors are usually inhibited by cell-cell contact. The absence of merlin favors their activation, stimulating mitogenic and survival pathways and favoring a lack of cell polarization. These alterations cause schwannoma cells to be unable to bind to an axon.²

The clinical differential diagnosis includes those lesions that present as a well-encapsulated tumor: granular cell tumor, salivary gland tumor, leiomyoma, neurofibroma, lymphangioma, hemangioma, cyst, lipoma, and others.

Treatment is surgical and is more complex in those tumors situated on the base of the tongue.⁶ Malignant change and recurrence are rare.²

We have presented this case as schwannoma is uncommon on the mucosa of the tongue but it is included in the differential diagnosis of benign tumors of the oral mucosa. As occurs with many of these tumors, the final diagnosis is made on histology of the surgical specimen.

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Fragmented Health Care Delivery in Ichthyosis[☆]



Dispersión en la atención médica de los pacientes con ictiosis

Congenital ichthyosis is a very rare genetic disease—an estimated 294 patients in Spain are thought to be affected.¹ The disease usually has a large impact on patients' quality of life because, regardless of the type of ichthyosis, patients may experience intractable pruritus, hyperhidrosis, failure to thrive during childhood, cicatricial alopecia, repeated keratitis, conductive hearing loss, and social problems in addition to cutaneous manifestations. As a result, in addition to dermatologists, most patients need to be attended by other specialists such as ophthalmologists, ear-nose-throat specialists, endocrinologists, nutrition experts, and psychologists. If it is rare for many dermatologists to see a patient with ichthyosis, it is easy to imagine the limited experience that many of these other specialists have.

During the first quarter of 2014, we conducted a survey of the members of the Spanish Ichthyosis Association (abbreviated as ASIC in Spanish) to find out the name of the attending physicians. Our aim was to determine whether dermatologists, ophthalmologists, ear-nose-throat specialists, and endocrinologists had experience in ichthyosis. Seventy-two ASIC members responded; in addition, ASIC has data on the dermatologists who attended a further 35 patients. In total, the ASIC members mentioned 70 different dermatologists. Of these, only 3 were mentioned by more than 5 different patients, 5 were the regular

dermatologist for 3 or 4 patients, 9 saw at most 2 patients regularly, and the remaining dermatologists (up to a total of 53) were mentioned by a single patient. A similar picture emerged with the other specialists, who were mentioned in the survey by at least half of the patients: 4 ophthalmologists were mentioned by 2 different patients, only 1 ear-nose-throat specialist was mentioned by more than 1 patient, and no endocrinologist of the 6 mentioned had more than one patient with ichthyosis (Table 1). Interestingly, several members highlighted that although they regularly attended appointments with these specialists, they were not usually seen by the same physician. In summary, our survey shows that very few physicians have experience with a significant number of patients with ichthyosis, and that it is likely that most patients are not attended regularly by other specialists who could help them with some of their possible comorbidities.

It is important to highlight possible biases in our survey. On the one hand, not all patients with ichthyosis belong to the ASIC (in fact, we have the names of dermatologists who attend approximately one third of all Spanish patients), and so the data only partially reflect the true situation. On the other, it is logical to think that dermatologists who recommend joining the ASIC are those who are more strongly represented in the survey. In any case, the proportion of dermatologists, ophthalmologists, and ear-nose-throat specialists who attended a single patient is noteworthy and probably reflects the true situation: most patients with ichthyosis lack a multidisciplinary follow-up, and are also seen by specialists with few other similar patients.

Table 1 Proportion of Patients With Ichthyosis Who Were Attended by the Same Specialist, According to the April 2014 Survey of Members of the Spanish Ichthyosis Association (ASIC).

	Dermatologists n = 70	Ophthalmologists n = 27	Ear-Nose-Throat Specialists n = 19	Endocrinologists n = 6
Attend 7 patients	1	–	–	–
Attend 6 patients	1	–	–	–
Attend 5 patients	1	–	–	–
Attend 4 patients	3	–	–	–
Attend 3 patients	2	–	–	–
Attend 2 patients	9	4	2	–
Attend 1 patient	53	23	18	6
Total Patients	107 (72 + 35 ^a)	31/72	22/72	6/72

^a Members of the ASIC who did not respond to the survey but whose information was available.

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