

Onchocerciasis Diagnosed in Italy

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Sir,

Onchocerciasis is a helminthiasis caused by the nematode *Onchocerca volvulus*. In spite of the fact that it affects almost 18 million people, onchocerciasis is scarcely known by western dermatologists. We describe two cases of onchocerciasis, which we have recently observed in Italy.

CASE REPORTS

Case 1. A 28-year-old Senegalese man was admitted because of a nodule localized in the dorsum of the left foot. The patient stated that he was in good general health and that the nodule had been present for about 6 months. It was round, 6 cm in diameter, covered by scales and crusts (Fig. 1), mobile, parenchymatous-hard and asymptomatic. The nodule was surgically excised. Histopathological examination confirmed the clinical suspicion of onchocercoma. Laboratory and instrumental examinations, as well as specialists' examinations, were negative. The patient did not turn up for therapy.

Case 2. A 60-year-old Italian man, a missionary in the Congo, was admitted because of diffuse pruritus of some months' duration. The patient stated that he was in good general health and that he was not on any therapy. Dermatological examination revealed a diffusely erythematous, xerotic and lichenified skin, with numerous excoriations. In addition, a nodule was observed on the left shoulder. It was round, 4 cm in diameter, covered by smooth skin, mobile, parenchymatous-hard and asymptomatic (Fig. 2).

The nodule was surgically removed. Histopathological examination showed, in the hypodermis, a cystic cavity, surrounded by a fibrous capsule. Inside the cyst, it was possible to recognize sections of the female of

O. volvulus (Fig. 3). Laboratory and instrumental examinations, as well as specialists' examinations, were negative. The patient was not treated as he returned to the Congo soon after the diagnosis.

DISCUSSION

Onchocerciasis is present in three geographical areas: sub-Saharan Africa, particularly Atlantic countries from Senegal to Angola; Arabic Peninsula and Central and South America (Mexico, Guatemala, Columbia, Venezuela, Ecuador and Brazil). The number of people living in endemic areas who are exposed to the risk of onchocerciasis is more than 85 million. Imported cases have been observed in France (1–6), Holland (7, 8), Italy (9), Spain (10) and the USA (11–14).



Fig. 2. Onchocercoma of the left shoulder of case no. 2.



Fig. 1. Onchocercoma of the dorsum of the foot of case no. 1.

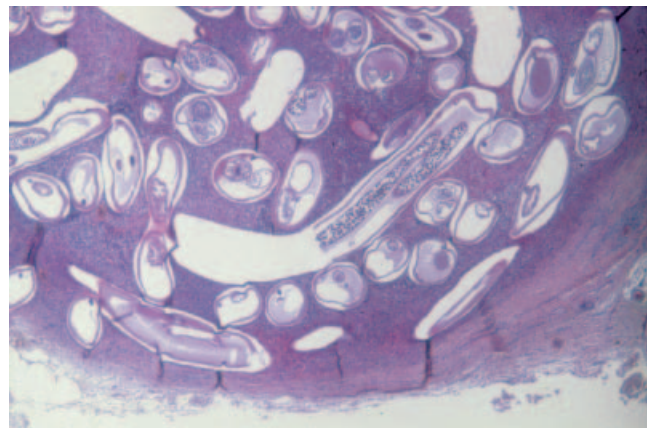


Fig. 3. Histology of a nodule from case no. 2 showing an onchocercoma with numerous cavities surrounded by a fibrous capsule.

The transmission of the disease occurs through the bite of *Diptera* females belonging to the genus *Simulium* (blackfly). In Africa, *Simulium damnosum* is particularly important. Blackflies are infected by biting an infected individual, especially during the day. Microfilariae are 'aspirated' by the vector directly from the dermis. Once microfilariae are penetrated, they develop into larvae. With subsequent bites, larvae are discharged from the buccal apparatus and, through a tiny break in the skin, they reach the dermis, subcutaneous tissue and lymphatics of a new host, where they mature into adult worms.

Adult worms live free or are massed together and surrounded by a fibrous capsule (onchocercoma). Fecundated females are able to produce hundreds of thousands of microfilariae, which hoard in dermis, subcutaneous tissue, lymphatics and lymph nodes. In these tissues, microfilariae move without periodicity. They can also reach the eyes, but they are not found in peripheral blood.

Skin involvement may be limited, even for a long period of time, to pruritus, which usually is diffuse and severe. It is due to the migration of microfilariae in the dermis and to the lysis of adult worms and microfilariae. Subsequent skin manifestations were classified by Murdoch et al. (15) as follows: I, Acute papular onchodermatitis; II, chronic papular onchodermatitis; III, lichenified onchodermatitis; IV, atrophy; V, depigmentation; VI, nodules (onchocercomas); VII, lymphadenopathy; VIII, hanging groin and IX, lymphoedema.

Onchocercomas represent the late phase of the disease. They are single or multiple (from 1–4 nodules up to, exceptionally, 100–150), with a diameter of 0.5–6 cm, usually covered by normal skin. Onchocercomas are mobile and parenchymatous-hard in consistency. Furthermore, they are asymptomatic and do not ulcerate, suppurate or calcify. They are often located around bony protruberances.

Laboratory tests show eosinophilia and increase in total IgE as well as inflammatory signs. The diagnosis of cutaneous onchocerciasis is based on the demonstration of microfilariae by means of the skin snip test: specimens of the skin are placed on a microplate with saline solution. After minutes or hours, the microfilariae, freed from the tissue, can be observed under the inversion microscope. The skin snip test is of limited value in early infestations and in cases with low microfilaridemia. Recently, PCR-ELISA has been successfully employed; it is a more sensitive test in cases with low microfilaridemia.

The histopathological picture varies according to the phase of the disease. Onchocercomas appear as round lesions, localized in the dermis and subcutaneous tissue, surrounded by a fibrous capsule, with multiple cavities

which are filled with adult worms and microfilariae. Around the capsule, lymphocytes, eosinophils, histiocytes, epithelioid cells and foreign body giant cells are present.

The therapy of onchocerciasis is based on ivermectin, although its mechanism of action is not known. It is active as a microfilaricidal agent as it rapidly and persistently reduces the number of microfilariae in the skin and eyes. Surgical excision of onchocercomas allows a reduction in the number of adult worms. Prophylaxis consists in the elimination of vectors. However, in the last few years, the development of resistance by some species of *Simulium* has been observed.

REFERENCES

1. Feuilhade de Chauvin M, Pouget F, Bernadou M, Revuz J, Touraine R. Un gros bras camerounais. *Ann Dermatol Venereol* 1986; 113: 968–971.
2. Ehram E, Thomas P, Hachulla E, Thomas P. Cas pour diagnostic. *Ann Dermatol Venereol* 1992; 119: 393–395.
3. Lanternier G, Le Guyadec T, Secchi T. Primo infestation onchocerquienne. *Nouv Dermatol* 1993; 12: 566–567.
4. Ochonisky S, Vignon-Pennamen MD, Fouchard N, Feuilhade De Chauvin M, Abirached G, et al. Onchocercose papuleuse aiguë localisée: une cause parasitaire du syndrome de Wells? *Ann Dermatol Venereol* 1995; 122(Suppl 1): S120–S121.
5. Le Guyadec T, Maccari F, Hernandez E, Gêrôme P, Le Vaguereuse R, Lanternier G. Cas pour diagnostic. *Ann Dermatol Venereol* 1999; 126: 729–731.
6. Lanternier G, Maccari F, Hernandez E, Le Guyadec T. Onchocercose de l'europeen de retour de séjour en zone d'endémie. *Nouv Dermatol* 1999; 18: 166–168.
7. Zuidema PJ. Onchocerciasis. *Dermatologica* 1977; 154: 54–56.
8. Van Den Hoogenband HM. Eosinophilic cellulitis as a result of onchocerciasis. *Clin Exp Dermatol* 1983; 8: 405–408.
9. Lombardo M, Girolomoni G, Pincelli C. Onchocercosi oculo-cutanea. Descrizione di un caso. *G Ital Dermatol Venereol* 1995; 128: 541–543.
10. Fonseca E, Del Pozo J, Del Castillo F, Contreras F, Cuevas J. Imported onchocerciasis in Spain. *J Eur Acad Dermatol Venereol* 1995; 5(Suppl 1): S174.
11. Maso MJ, Kapila R, Schwartz RA, Wiltz H, Kaminski ZC, Lambert WC. Cutaneous onchocerciasis. *Int J Dermatol* 1987; 26: 593–596.
12. Vernick W, Turner SE, Burov E, Telang GH. Onchocerciasis presenting with lower extremity, hypopigmented macules. *Cutis* 2000; 65: 293–297.
13. Borup LH, Peters JS, Sartori CR. Onchocerciasis (river blindness). *Cutis* 2003; 72: 297–302.
14. Okulicz JF, Stibich AS, Elston DM, Schwartz RA. Cutaneous onchocercoma. *Int J Dermatol* 2004; 43: 170–172.
15. Murdoch ME, Hay RJ, Mackenzie CD, Williams JF, Ghalib HW, Cousens S, et al. A clinical classification and grading system of the cutaneous changes in onchocerciasis. *Br J Dermatol* 1993; 129: 260–269.