

**Title:** Sixth nerve palsy associated with obstruction in Dorello's canal, accompanied by nodular type muscular sarcoidosis

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## **Introduction:**

Sarcoidosis is a chronic multisystem granulomatous disease [1]. Every organ can be affected and sarcoidosis can cause a variety of neurological impairments. The prevalence of clinical involvement of the nervous system is estimated to be 5–15% [2,3], but the prevalence of subclinical neurosarcoidosis may be much higher [4,5] and potentially lethal manifestation of sarcoidosis [1]. Cranial neuropathies are the most common manifestation of neurosarcoidosis [3]. Only three in 50 patients with neurosarcoidosis showed abducent nerve palsy [6], but the cause was not described. Dorello's canal, an osteofibrous canal at the apex of the petrous bone containing the abducent nerve, supposed to be important in suffering the abducent nerve, however there is no report presenting abducent nerve palsy related to the canal in neurosarcoidosis patients.

Here, we report a case of sarcoidosis with symptomatic abducent nerve palsy caused by the obstruction in Dorello's canal, and the palsy and the obstruction were improved by oral steroid therapy. Brain and muscle MRI findings, and the patient's muscle-biopsy confirmed our diagnosis.

## **Case Report:**

A 52-year-old Japanese woman presented with horizontal double vision for 10 days before hospitalization. Her left eye rotated inwardly when she looked straight ahead. She was examined by a neurologist and admitted to our hospital. She had no previous history of diplopia and there was no familial history of neuromuscular disease. Her general physical examination was unremarkable. Neurological examination confirmed left abducent nerve palsy. A manual muscular test revealed a normal score in the face, neck, and extremities. Muscle amyotrophy and myalgia were not noted, but the thigh muscles were slightly swollen. The deep tendon reflex was normal. No cerebellar or sensory abnormalities were observed. Her serum angiotensin-converting enzyme (ACE) was slightly elevated to 36.2 IU/L (normal, 7.7–29.4), and her lysozyme level was also increased to 11.7  $\mu\text{g/mL}$  (4.2–11.5). Thyroid function, erythrocyte sedimentation rate, rheumatoid factor, anti-nuclear antibody titer, serum immunoglobulin, and CK were normal. The results of all tests of the cerebrospinal fluid were normal. The tuberculin reaction was negative. Her chest X-ray and CT revealed mild enlargement of the bilateral mediastinal and hilar lymph nodes. Her brain MRI showed obstruction in the spinal fluid space of the left Dorello's canal (Fig. A). The muscular MRI of her thighs revealed a star-shaped central structure of decreased signal intensity on axial images (Fig. C; arrowheads), and a long nodule with an inner stripe of decreased signal

intensity and outer stripes of increased signal intensity on the coronal and sagittal images (Fig. D; arrows). The former finding is called the “star-shaped sign” and the latter is the “three stripes sign”. These are consistent with nodular-type muscular sarcoidosis [7]. Muscle-biopsy specimens from her quadriceps femoralis contained granulomatous epithelioid giant cells and non-necrotizing chronic lymphadenitis, which are also consistent with sarcoidosis. Based on these findings, it was considered that the obstruction of the abducent nerve by granulomas in Dorello’s canal was the cause of her left abducent palsy. She was treated with oral prednisolone (1 mg/[kg body weight]) for 30 days and the dose was gradually reduced. Twenty-one days later, the abducent nerve palsy was fully improved and the fuzziness of the left Dorello’s canal became clear (Fig. B). Her clinical improvements were also associated with a decrease in serum ACE to the normal range. Oral prednisolone was reduced to 8 mg per day within 3 years, and she experienced no recurrence.

### **Discussion:**

The abducent nerve passes a long distance from the brainstem to the lateral rectus muscle.

There are three segments in the course of the abducent nerve, the subarachnoid segment, the petroclival segment, and the intracavernous segment [8]. Dorello’s canal is a part of the

passageway of the abducent nerve, which is in the petroclival segment. In our case, brain MRI showed the obstruction of the spinal fluid space in the left Dorello's canal. Other spaces where the abducent nerve passed, including the cavernous sinus, were clear. The cause of the abducent nerve palsy in our case was believed to be compression by sarcoid granulomas in the left Dorello's canal, because the obstruction was reduced by prednisolone and the symptoms simultaneously resolved. There is only one case report presenting abducent nerve palsy related to Dorello's canal, but this was due to hypoplasia of the canal [9]. The present case is the first that showed abducent nerve palsy due to obstruction in Dorello's canal. The combination of nodular-type muscular sarcoidosis is also a characteristic in this case. Symptomatic muscle sarcoidosis is rare and has only been described in 0.5%–2.3% of sarcoidosis patients [10]. In nodular-type sarcoidosis, MRI findings can provide specificity for this condition [7]. The nodule is divided into two structures, central and peripheral, that form “dark star sign” and “three stripe sign”. Histopathologically, the central structure consists of dense connective tissue and hyaline material, presumably due to chronic inflammation and very low cellularity. The peripheral structure of the nodule consists of the high cellularity of active inflammatory granulomas and edema [7]. In our case, only tension in the thigh was recognized on the physical examination, and no

other muscle symptoms were observed. However, the characteristic signs in muscular MRI were useful for diagnosis.

Thus, we present a case of sarcoidosis, which showed cranial neuropathy and nodular-type muscular sarcoidosis. If abducent nerve palsy is detected, it is important to consider sarcoidosis and compression in Dorello's canal as its cause. It is also important to note that muscle MRI findings are useful for the diagnosis of muscular sarcoidosis.

## **Legends**

Figure. (A) and (B) On a thin slice-enhanced T1-weighted reconstruction MRI, the spinal fluid space within the left Dorello's canal (arrowheads) was fuzzier than that in the right Dorello's canal (arrows) at disease onset (A). After steroid therapy, the fuzziness in the left Dorello's canal was reduced (B, arrowheads). (C) T2-weighted reconstruction MRI showed an inner stripe of decreased signal intensity and outer stripes of increased signal intensity ("three stripes sign") (arrowheads). (D) The short tau inversion recovery (STIR) reconstruction MRI images showed a star-shaped central structure of decreased signal intensity ("dark star sign") (arrowheads).

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