Paraneoplastic retinopathy associated with retroperitoneal liposarcoma

Abstract: We report a case of paraneoplastic retinopathy associated with a retroperitoneal liposarcoma. A 42-year-old man was referred to our hospital with complaints of night blindness and blurred vision in the peripheral field. Electoretinograms showed a progressive amplitude reduction in his both eyes. Abdominal magnetic resonance imaging showed a large retroperitoneal mass, and pathologic examination revealed a dedifferentiated liposarcoma. Western blot analysis showed an antiretinal antibody in the serum of our patient, and his serum reacted with the photoreceptors of a bovine retina. To the best of our knowledge, this is the first case of paraneoplastic retinopathy associated with a liposarcoma.

Keywords: paraneoplastic retinopathy, retroperitoneal liposarcoma, electoretinogram, cancer-associated retinopathy

Introduction
Paraneoplastic retinopathy (PR) is a progressive retinal disease caused by antibodies generated from neoplasms distant from the eye.1–3 The retinopathy can develop either before or after the diagnosis of the neoplasm. Patients with PR usually have night blindness, photopsia, ring scotoma, attenuated retinal arteriole, and abnormal electoretinograms (ERGs). PR is thought to be mediated by an autoimmune mechanism, and is associated with the presence of antiretinal autoantibodies in the serum.

Various types of neoplasms are known to cause PR, including malignancies of the lung, breast, cervix, colon, prostate/bladder, uterus/endometrium, and blood cells. Only two cases of PR associated with a sarcoma, a malignant tumor arising from mesenchymal cells, have been reported.4,5 We report a case of PR associated with a retroperitoneal liposarcoma. The patient’s visual symptoms preceded the discovery of the tumor by six months.

Case report
A 42-year-old man was referred to our hospital with complaints of night blindness and blurred vision in the peripheral field. He did not have any systemic or eye diseases including a malignant tumor, and the family history revealed no other members to have any eye diseases.

At the initial examination, his best-corrected visual acuity was 1.0 in both eyes, but Goldmann perimetry showed defects in the mid-peripheral visual fields in both eyes (Figure 1A). Ophthalmoscopy showed that the fundus was nearly normal, but fluorescein angiography demonstrated mottled hyperfluorescence along the vascular...
ardes (Figure 1B and 1C). The ERG amplitudes of both the rod and cone components were reduced (Figure 1D, middle column). Based on these findings, we diagnosed him as having a rod-cone dystrophy.

However, his symptoms progressively worsened, and the amplitudes of the ERGs were further reduced six months after the initial examination (Figure 1D, right column). We then suspected PR, and performed systematic magnetic resonance imaging (MRI). The abdominal MRI showed a large retroperitoneal mass (Figure 2A, arrow) which compressed the left kidney.

We also performed Western blot analysis using bovine retinal proteins to determine whether there were any antiretinal antibodies in the serum of our patient. A retinal protein of approximately 83 kD (Figure 2B, arrow) was detected in the serum of this patient. We also confirmed that the serum reacted with the photoreceptors of a bovine retina (Figure 2C).

We then diagnosed our patient as having PR associated with retroperitoneal tumor, and the tumor as well as the left kidney was removed (Figure 2D). Pathologic examination revealed a dedifferentiated liposarcoma that contained the characteristic two patterns of a well differentiated liposarcoma (Figure 2E, asterisk) and dedifferentiated fibrotic sarcomatoid tissue (Figure 2E, arrow). After the tumor was resected, he received chemotherapy but he had a recurrence with metastasis. His status became unknown after he moved to his hometown for terminal care.

**Comments**

A PubMed search for cases of PR associated with a sarcoma yielded two cases.4,5 One case involved a uterine sarcoma, and the other a rhabdomyosarcoma of the thorax. To the best of our knowledge, this is the first case of PR associated with a liposarcoma. A liposarcoma is
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a malignancy of fat cells that occurs in deep soft tissue
and is mostly seen in the limbs and retroperitoneum.5 It
is the most common soft tissue sarcoma and accounts for
approximately 20% of all mesenchymal tumors. Most of
the patients with liposarcoma have no symptoms until
the tumor becomes large and causes pain or functional
disturbances in neighboring organs.

We detected an antiretinal antibody in the serum of
our patient, and found that the serum reacted with the
photoreceptors of a bovine retina, suggesting that this anti-
body caused the retinopathy of our patient. However, we
did not confirm that this antibody actually reacted to the
tumor proteins of our patient. Thus, additional experiments
are needed because it is known that the antiretinal antibody
can be produced not only in PR, but also in other retinal
degenerative diseases as a secondary complication of retinal
cell death.7

Our experience with this case demonstrated that it is
important for ophthalmologists to be aware that liposarcoma
can be the cause of PR. In these cases, the visual symptoms
may precede the discovery of this tumor, because liposarcoma
usually grows silently in deep soft tissues without any local
symptoms.

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Disclosures

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