
Anahtar Kelimeler
En Coup De Sabre; Skleroderma

*This study was presented as an e-poster in 12. Aegean Dermatology Days, held in Bodrum between May 10th-14th.
Introduction
Morphea (localized scleroderma) is an inflammatory skin disease that causes sclerosis in the dermis and subcutaneous adipose tissue [1]. Linear scleroderma is the most common form of localized scleroderma in children and adults. The term en coup de sabre (ECDS) is used to describe a variant form of linear scleroderma, which has a special appearance in the frontal or frontoparietal region. ECDS emerges in the form of linear, erythematous bands, and gradually transforms into firm plaques. It is usually seen on the paramedian region on the forehead and is unilateral [2]. Here, we present a case of an 18-year-old female patient, who is diagnosed with ECDS after clinical and histological examinations.

Case Report
An 18-year-old female patient was admitted to our clinic with collapse and discoloration on the forehead line. The patient’s medical history showed that pink-purple color change started approximately four years ago, which was followed by a progressive collapse on the forehead line. The patient’s family history did not show anyone with a similar disease. Systemic examination was normal, and physical examination did not show any pathological findings. Dermatologic examination showed that the patient had an atrophic, linear, and depressed lesion on the frontal region extending to the scalp [Figures 1 and 2]. The patient’s routine laboratory tests results were within expected intervals. The patient underwent skin biopsy from the frontal region. A written informed consent form was obtained before the procedure.

Histopathological examination showed superficial orthokeratosis and irregular acanthosis in the epidermis. An increase in the number of thick, homogenized, collagen fibers, involving the whole dermis and affecting the subcutaneous adipose tissue, and intermittent atrophy in skin adnexa were present [Figure 3]. Verhoeff-Van Gieson staining (VVG) showed thick, fragmented, elastic fibers laying parallel to the dermis surface [Figure 4].

Discussion
ECDS is a rare variant of morphea. The disease is usually seen during childhood, and the average age at onset is ten years [2]. A retrospective study on 82 patients indicated that ECDS has an incidence of 0.13/100,000 [3].

Similar to other types of scleroderma, the exact etiology of ECDS remains unknown [2,4]. The disease is considered to have an autoimmune origin and triggered by environmental factors. The exact relationship between progressive facial hemiatrophy (Parry Romberg Syndrome), which is characterized by progressive atrophy on one half of the face, and ECDS is not known. Still, given the similarities in the pathogenesis of these diseases, they are considered to lie on different ends of the spectrum of the same disease [5]. In the present study, our patient did not have facial atrophy.

Neurological involvement with ECDS has been identified in 18-47% of the cases. Epileptic seizures are the most common symptoms, while hemiparesis, muscle weakness, headache, personality changes, and deterioration in intellectual functions have also been reported [6,7]. Our patient’s neurological examination was normal, and cranial MRI did not reveal any pathological findings. It is also possible to detect ophthalmologic anomalies in ECDS; however, our patient did not have any.
abnormal findings in the ophthalmic examination. Methotrexate and systemic glucocorticoids represent the first line treatment for ECDS, and it is recommended to maintain treatment with UVA1, PUVA phototherapy, narrowband UVB or mycophenolate mofetil. Other treatment options include topical tacrolimus, topical vitamin D and glucocorticoid combinations, and imiquimod [8].

In conclusion, we wanted to present this case as ECDS represents a rare variant of morphea.

Ethical Responsibilities: All institutional and national guidelines for the care and use of laboratory animals were followed. Conflict of Interest: No potential conflict of interest relevant to this article was reported. Funding: The funders had no role in study design, data collection, and analysis, decision to publish, or preparation of the manuscript.

Competing interests
The authors declare that they have no competing interests.

References

How to cite this article: