

Bag of Worms in Orbital/Periorbital Plexiform Neurofibroma, from the Macroscopic to the Microscopic View

González-Valdivia H^{1*}, Figueroa-Vercelino J¹, Casas-Gimeno E¹, Suñol-Capella M² and Prat-Bartomeu J¹

¹Department of Ophthalmology, Sant Joan De Déu Hospital, Barcelona, Spain

²Department of Anatomical Pathology, Sant Joan De Déu Hospital, Barcelona, Spain

*Corresponding Author: González-Valdivia H, Department of Ophthalmology, Sant Joan De Déu Hospital, Barcelona, Spain.

Received: February 26, 2021

Published: March 18, 2021

© All rights are reserved by González-Valdivia H., et al.

Neurofibromatosis 1 (NF1) is a neurocutaneous disorder with autosomal dominant inheritance caused by genetic alterations of the NF1 gene [1,2]. About 30% of NF1 patients develop plexiform neurofibromas (PNFs), a benign tumor of the peripheral nerve sheath which originates from subcutaneous or visceral nerves [4]. When PNFs affect the periorbital zone in children, both vision and eyelid functionality can be compromised and surgery may be necessary to prevent amblyopia or protect the cornea [2,3]. We present anatomical and histological images of an orbitopalpebral plexiform neurofibroma in a pediatric patient who had the typical sign of bag of worms before debulking surgery [4].



Figure 1: Debulking surgery: Anatomic appearance of the plexiform neurofibroma.

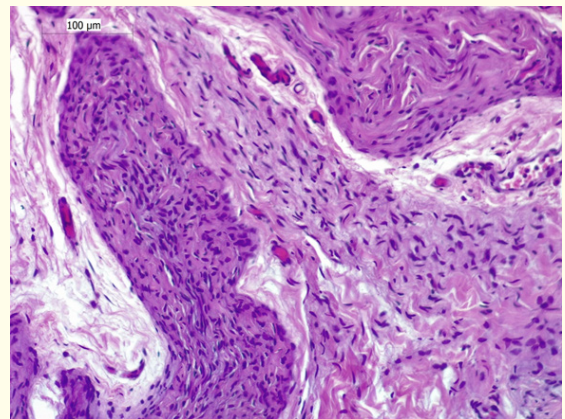


Figure 2: Histopathological findings: Tumor cells elongated on a myxoid background, distributed adopting a plexiform pattern.

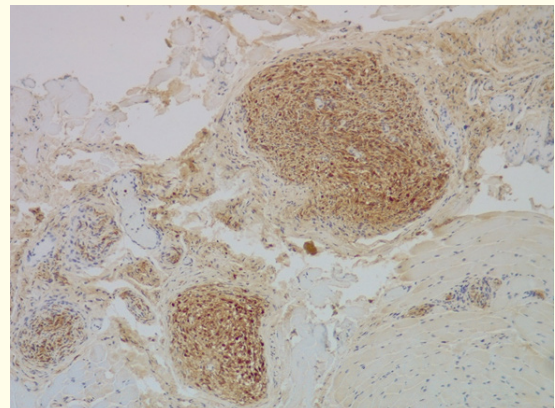


Figure 3: Histopathological findings: Positivity of tumor cells for S100 protein in immunohistochemistry.

Bibliography

1. Ly Ky and Blakeley JO. "Diagnosis and Management of Neurofibromatosis Type 1". *Medical Clinics of North America* 103.6 (2019): 1035-1054.
2. Avery RA., et al. "Orbital/Peri-Orbital Plexiform Neurofibromas in Children with Neurofibromatosis type 1: Multi-disciplinary Recommendations for Care". *Ophthalmology* 124.1 (2017): 123-132.
3. Avery RA., et al. "Visual outcomes in children with neurofibromatosis type 1 and orbitotemporal plexiform neurofibromas". *American Journal of Ophthalmology* 155 (2013): 1089-1094.
4. Patil K., et al. "Facial plexiform neurofibroma in a child with neurofibromatosis type I: a case report". *Journal of Indian Society of Pedodontics and Preventive Dentistry* 25.1 (2007): 30-35.

Assets from publication with us

- Prompt Acknowledgement after receiving the article
- Thorough Double blinded peer review
- Rapid Publication
- Issue of Publication Certificate
- High visibility of your Published work

Website: www.actascientific.com/

Submit Article: www.actascientific.com/submission.php

Email us: editor@actascientific.com

Contact us: +91 9182824667