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Neurosurgeon's role in neurocutaneous syndromes

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Neurocutaneous Syndromes are a heterogeneous and complex group of genetic disorders characterized by dermatologic, ophthalmologic and neurological findings, which include over 40 different entities. The same molecular defects lead to skin and CNS alterations, and predispose patients to a significant risk of neoplastic lesions. CNS tumors in phakomatoses differ from sporadically occurring version of the same tumors, in natural history, histological features and prognosis. Among patients with neurocutaneous syndromes exists a great complexity and variability in genetic expression and clinical course, which reflect the opportunity, either, to categorize the patients suffering from the same syndrome into different subgroups, and to establish the better treatment's strategy. We performed a literature's review, in the light of the cases evaluated and treated at the Neurosurgical Clinic of the University of Messina, Italy, focusing on the role of neurosurgeon in neurocutaneous syndromes to underline who, when and what to treat, to understand if his role is, to date, clearly defined and to discuss if some challenges should be needed.