Soft tissue sarcoma complicating neurofibromatosis type 1 during pregnancy

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- **Introduction :**

Neurofibromatosis type 1 (NF1) is the most common genodermatosis. This autosomal dominant disease predisposes to the development of benign tumors (neurofibromas), but also malignant tumors [1]. Soft tissue sarcomas (STS) are rare tumors characterized by anatomic, histological and prognostic heterogeneity. Genetic predisposition participates in 3% of the STS and corresponds essentially to the NF1 [2]. We report the case of 24 year old female with NF1 who has developed a soft tissue sarcoma during her first pregnancy .

- **Case report :**

It is a 24-year-old patient with NF1, who developed a mass of the right flank during her first pregnancy, rapidly increasing in size, discovered in postpartum (fig 1) . Thoraco-abdominopelvic CT showed in the right flank a parietal mass of heterogeneous density infiltrating the inferior vena cava, with close contact with the ipsilateral iliac bone with no other lesions (fig 2) . An excision of the tumor was performed witch histological examination showed an undifferentiated sarcomatous process with muscle infiltration and invasion of the bone cortex. The decision was to complete by radiochemotherapy with close monitoring .

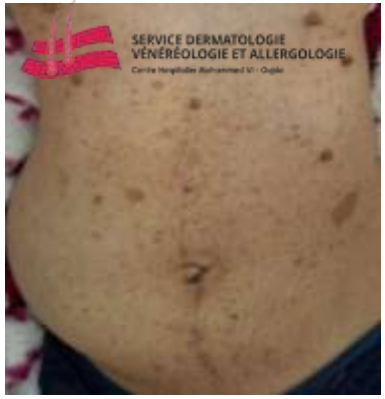

Fig 1 : Mass of the right flank

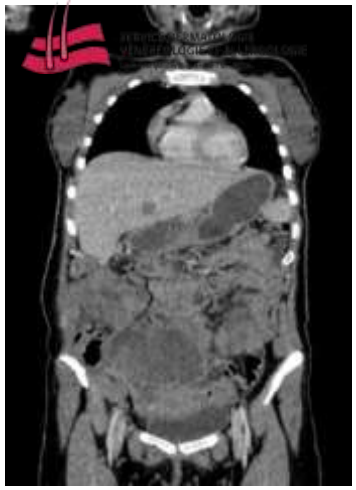

Fig 2 : Thoraco-abdominopelvic CT showing parietal mass of the right flank

- **Discussion :**

NF1 is a frequent genetic disease that affects from 1/4000 to 1/3000 individuals with a homogeneous global distribution in the world [3]. Its prognosis and its genetic mechanisms have been the subject of many publications for the last fifteen years [4]. However few studies have investigated the prevalence and impact of malignancies in NF1 [1]. Patients with NF1 are four times more likely to develop a tumoral pathology [5]. Frequently observed tumors are melanoma, pheochromocytoma, medullary thyroid carcinoma, glioma of the optic pathway, leukemia and breast cancer [6]. Sarcomatous transformation is exceptional [2]. The main risk factors are the presence of isolated cutaneous, visceral, plexiform and / or nodular neurofibromas, witch require careful monitoring in these patients [7]. Some studies have tried to establish the link between STS and pregnancy, and have shown an increased risk of STS with late age at first pregnancy [8].

For a better understanding of such a case, many questions are to be asked : Is it sarcoma newly emerged during pregnancy or visceral neurofibroma transformed into sarcoma ? Does the development of this tumor during pregnancy remain a coincidence? Or is pregnancy a new risk factor for the development of these tumors?

- **Conclusion :**

The development of malignancies during NF1 emphasizes the importance of prevention counseling as well as the regular monitoring of patients with this disease, especially patients with risk factors or alarming signs [1].

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