Meningioma and melanoma in patients with Turner syndrome  R.J.W De Keizer MD PhD; N. Lauwers MD; V. De Groot MD PhD FEBO Antwerp University and University Hospital Antwerp Belgium.

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Purpose: to highlight the increased risk of uveal melanoma and meningioma in Turner syndrome.

Methods: We report 2 cases: one with an uveal melanoma, one with sphenoid meningioma and conducted a literature search.

Results: A 43 years old female with Turner syndrome was enucleated because of a large uveal melanoma. Genetic analyses showed anomalies in chromosome 8. A literature study did not revealed any overlap with the 45X/46 XY, partial /complete Turner syndrome. A study of 3425 Turner patients revealed an increased risk for eye/orbital tumors (standard incidence ratio 10.5 and cerebral meningioma ( SD ratio of 12.0(1), confirmed by Pier.(2). Female 2 presented with bilaterally multiple meningomas(middle fossa, parasellar, sphenoid). Neurosurgery was performed in 1994 and 2007 with irradiation. The Left Eye was blind, and the RE was deteriorating. Neither steroids nor Nolvadex did stop the progression. Since the tumor was positive for progesterone receptors, we suggested a progesterone antagonist. Unfortunately complex surgery was performed after which the patient deceased. Turner patients require live long hormonal replacement starting in early puberty,with estrogens and progesterone’s. Growth of meningiomas can be induced by female hormones, and Turner patients have more melanocytic nevi.

Conclusion: Turner syndrome could be associated with an increased risk of melanoma and meningiomas. Whether their live-long hormonal substitution might be a causal factor, still remains to be determined. Regular ophthalmic check-up is necessary. If progression of the tumor is detected, multidisciplinary approach is mandatory.

1) Schoemaker M.J. Lancet oncology 2008,9-239

2).Pier D.P Eur j Gen 2014,57.269-274