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Fibroadenoma of the Breasts and Ovarium in a 16yo Girl with EMG Syndrome

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[Introduction] Beckwith-Wiedemann syndrome (BWS) has an increased risk of developing childhood neoplasms. However, the span of tumorigenicity is not well documented. The authors report a female case of BWS, which presented fibroadenoma arising from bilateral breasts and the right ovary at the age of 16 years. This is the oldest case of BWS that presented tumorigenesis reported in the English literature.

[Case Report] A female baby was born with BWS; gigantism (birth weight +5.7SD), omphalocele, macroglossia and nesidioblastoma (NB). On the first day of her life, a radical operation was performed for omphalocele, concomitantly with hepatic cyst. For the NB, a subtotal pancreatectomy was performed at the age of 8 years. At 16-year-old, she presented breast tumors, the biggest one was resected and the others were sampled, and histologically diagnosed as fibroadenoma (FA). Three months later, she presented an ovarian tumor, which was removed laparoscopically and histological study revealed FA. Her two years’ postoperative course was uneventful.

[Discussion] Until now only one case of BWS, which presented FA at one-year-old, had been reported. Although it is not clear whether the tumors occurred as a consequence of BWS or coincidentally, this case poses the question of the duration of tumorigenicity.