

Table 1 - Details of clinical features of patients 1 and 2

	Skin findings	CNS findings	Bone findings	Endocrinological findings	Management
Patient 1-1	<p>Large right sided Blaschkolinear KEN (scalp, face, neck, thorax, abdomen and back). Woolly hair naevus adjacent to KEN.</p>		<p>Osteofibrous dysplasia of right lower tibia</p>	<p>Normal serum calcium, phosphate, alkaline phosphatase, Vitamin D3 (low normal limit) and Parathormone FGF-23 128 RU/mL (normal \leq 125). Right sided ovarian cyst.</p>	<p>8 years: Presented with pain in right leg. Vitamin D started. 13 years: curettage of cyst in right lower tibia and bone transplant from hip. 15 years : Crutches and wheelchair bound. Excision with transplant and fixation with intramedullary pin, because of persistent pain.</p>
Patient 1-2	<p>Phakomatosis pigmentokeratolica: - KEN left scalp, face, neck, lip, palate and tongue. - Papular naevus spilus in checkerboard pattern: right side of upper back, left shoulder, both ankles and the left forearm - Perioral squamous papilloma.</p>	<p>MRI brain: small anterior pituitary</p>	<p>Osteofibrous dysplasia of left mid tibia with anterior bowing, progressive after surgery with poor remodeling. After 2 surgical procedures still requires crutches and wheelchair.</p>	<p>Normal serum calcium, phosphate and FGF-23 Growth hormone deficiency secondary to neurosecretory dysfunction thought to be connected to small anterior pituitary.</p>	<p>Bone dysplasia: Infancy: Splint lower left leg. 4 years: osteotomies and insertion of Fassier-duval intramedullary nail. 8 years: revision intramedullary rush pin left tibia, proximal tibial osteotomy, excision of 5 cm left diaphysis and application of a lizarov external fixator. Vitamin D supplementation from 8y.</p>