

# TOURNAINE-SOLENTE-GOLE SYNDROME:THE COMPLETE FORM

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## INTRODUCTION

Primary or familial hypertrophic osteoarthropathy also referred to as pachydermoperiostosis (PDP) or touraine solente gole syndrome.Autosomal dominant pattern of inheritance. M:F 9:1.

## CASE SUMMARY

45/M presented with pandigital clubbing,thickened greasy &coarse skin,furrowed forehead with ptosis,palmoplantar hyperhidrosis,generalised seborrhic dermatitis& multiple folliculitis in both lower limbs & dyspepsia.

## IMAGING

X-Ray both hands&feet: Shows soft tissue thickening noted in all fingers & toes with e/o irregular periosteal thickening noted in metacarpals, metatarsals & proximal phalanges of both feet & hands.

X-Ray both forearm & legs:Irregular periosteal proliferation with cortical thickening of both radius, ulna & both tibia &fibula .E/O interosseous membrane ossification noted in Lforearm.

## <sup>99m</sup>Tc-METHYLDIPHOSPHONATE BONE SCAN

Pericortical linear accumulation of radiotracer along the shaft of long bones & proximal phalanges .

## UPPER GI ENDOSCOPY

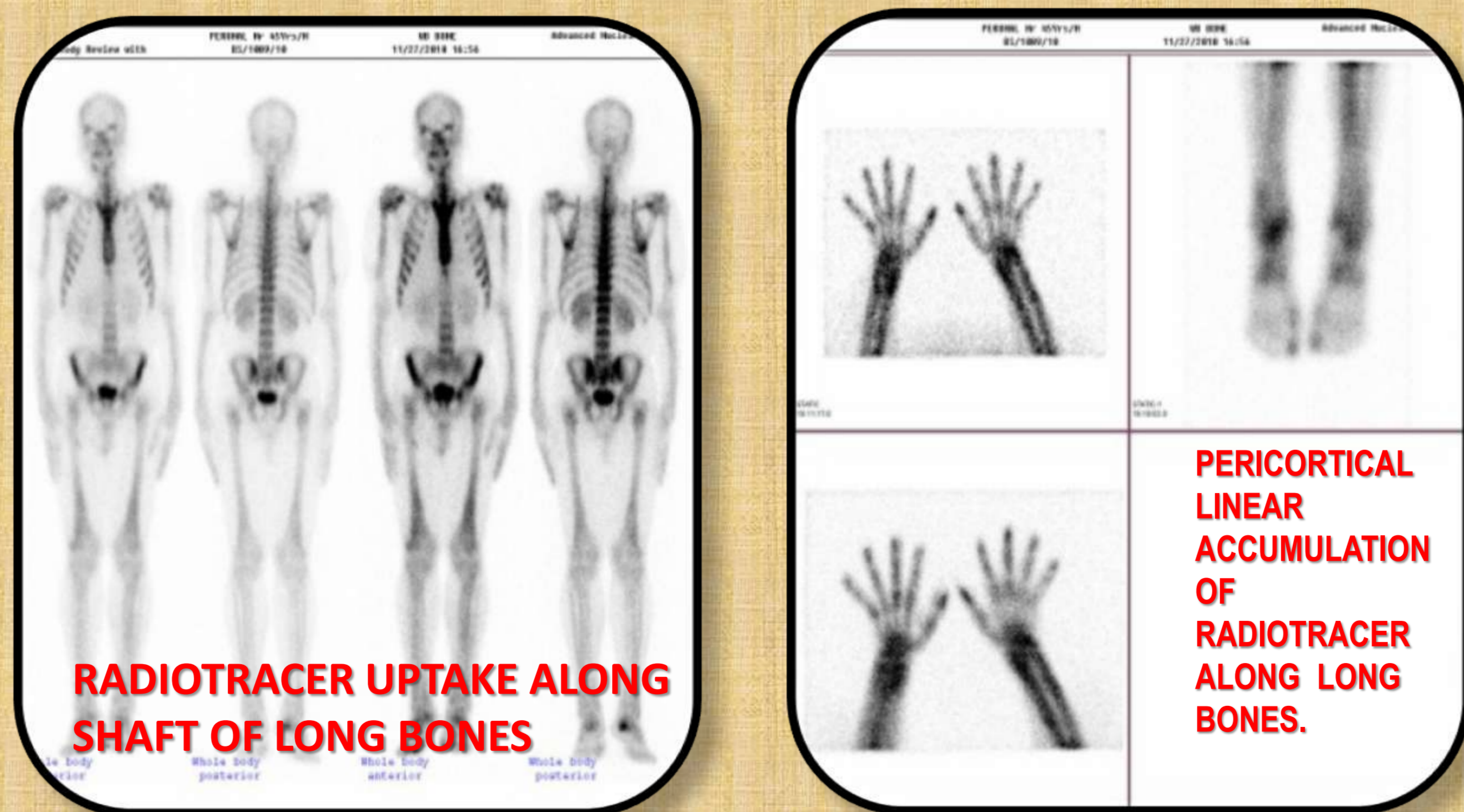
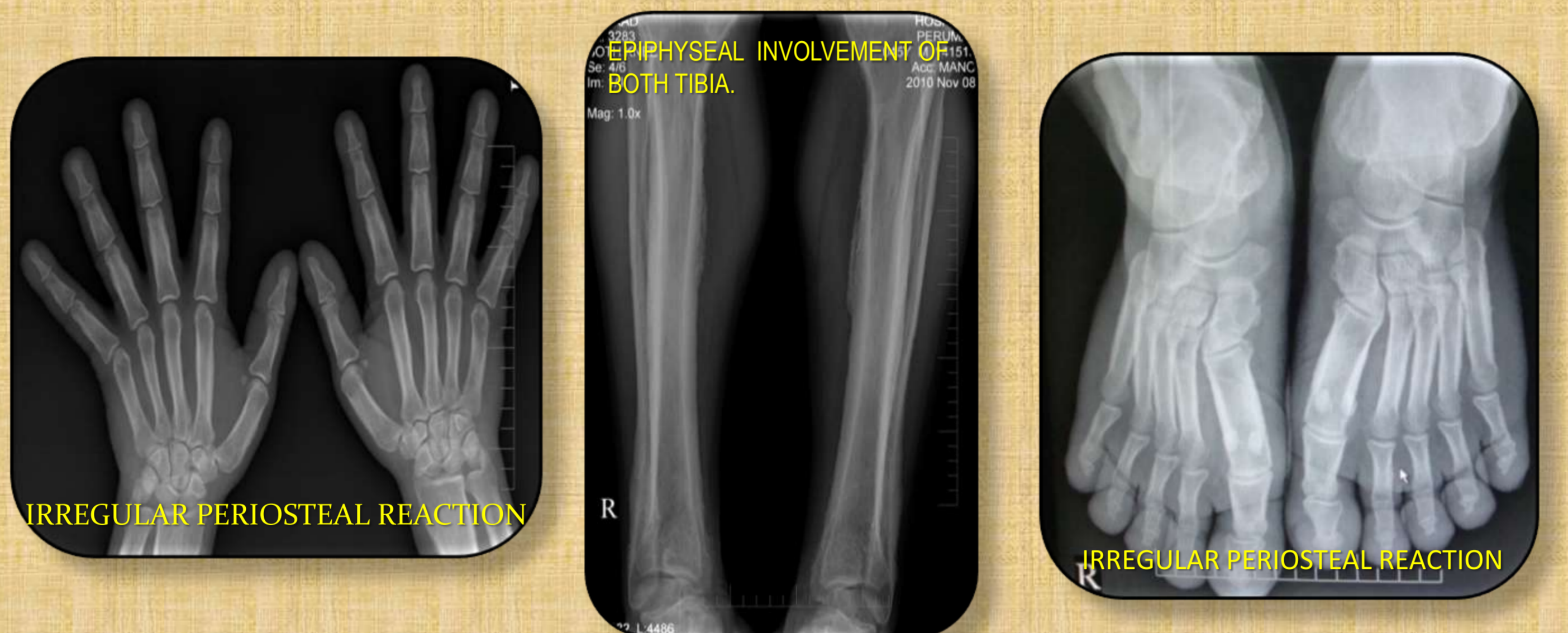
Prominent gastric mucosal folds with nodularity noted in entire stomach.

## CT ABDOMEN WITH IV & NEUTRAL CONTRAST

E/O uniform wall thickening (>1 cm) noted in the entire stomach & pylorus.

## MICROSCOPY

E/O thickening& packing of collagen fibers in dermis with hypertrophy of epidermal appendages with seborrhic hyperplasia-suggestive of pachydermia.



Primary osteoarthropathy has to be differentiated from the secondary form .

FEATURES	PRIMARY	SECONDARY
AGE OF ONSET	Starts earlier	later
COURSE	Remains static	progressive
SKIN MANIFESTATION	Prominent	Less common
JOINT PAIN	Minimum	Severe
EPIPHYSEAL INVOLEMENT	Yes	No
UNDERLYING CAUSE	None	Ass. With bronchogenic CA,Bronchiectasis etc

## DIAGNOSIS

Having excluded the secondary causes & with typical clinikoradiological features the diagnosis of TOURAINE-SOLENTE-GOLE SYNDROME with HYPERTROPHIC GASTROPATHY IS MADE.

## DISCUSSION

PDP is a rare hereditary disorder being classified into

- Complete form-pachydermia,clubbing,periostosis .
- Fruste form-prominent pachydermia with minimal skeletal changes.
- Incomplete form-no pachydermia.

## COMPLICATIONS

May develop severe kyphosis, restricted motion, and neurologic manifestations.

## DIFFERENTIAL DIAGNOSIS

Acromegaly,Thyroid acropachy,Syphilitic periostosis.

## TREATMENT&FOLLOW UP

NSAIDS,Colchicine,Pamidronate,Tamoxifen citrate & Isotretinoin. Progression typically caeses after 10 years.

## PATIENT EDUCATION

Genetic counselling to be offered to patients & their families.

## REFERENCES

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