Case Presentation – 1

…..*A pain in the neck*

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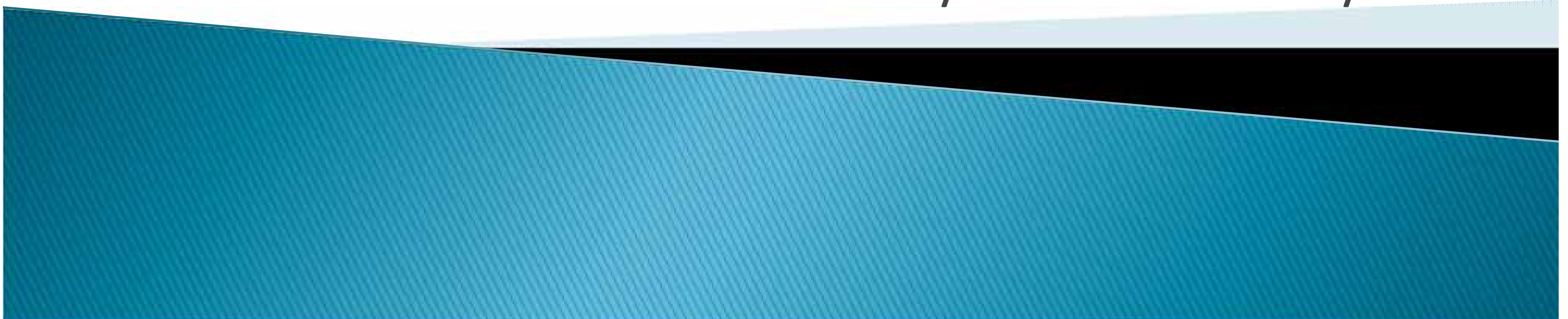


History

- I.D - 12 years - Female
- Headache since 07.08.13
- Fever, vomiting, neck pain started 09.08.13
- Attended govt hosp – prescribed injection/tablets + amoxil
- No improvement – Attended govt hosp again 2 days later – injection – vomiting settled

History

- Persistent fever, neck pain and aching upper arm
- Admitted JH 16.08.13 – Ix done – all normal – Discharged next day on amoxil + 3 tablets ?which
- Diplopia on day of discharge
- Presented to ABH ED 19.08.13
- Exam: Neck stiffness ++++; diplopia; No squint or nystagmus. No cerebellar signs. VF normal.

Differential diagnosis

- Partially treated bacterial meningitis
- Viral meningitis
- Encephalitis
- Brain abscess
- Lyme disease
- SOL

Management – 1

- Iv ceftriaxone – high dose
- MRI brain: normal
- TLC 15.2; CRP 2.4; ESR 28
- LP under GA

CSF

	Protein mg/dL	Glucose CSF:Blood ratio	WBCs (/microL)
Normal	20-45	>50%	<4 (1-3% N)
Patient	**21**	**70%**	**95 (13,400 RBCs). N 2%**
Bacterial meningitis	100-500	<40%	100-60,000+ (N mostly)
Partially treated meningitis	100+	↓ or Normal	1-10,000 (mixed N/L)
Viral meningitis/ encephalitis	20-100	Normal	<1000 (N early, L later)
Brain abscess	20-200	Normal	0-100 (N)
Traumatic tap	1mg/dL for every 1000 RBCs		1 WBC for every 500-700 RBCs

Ophthalmology

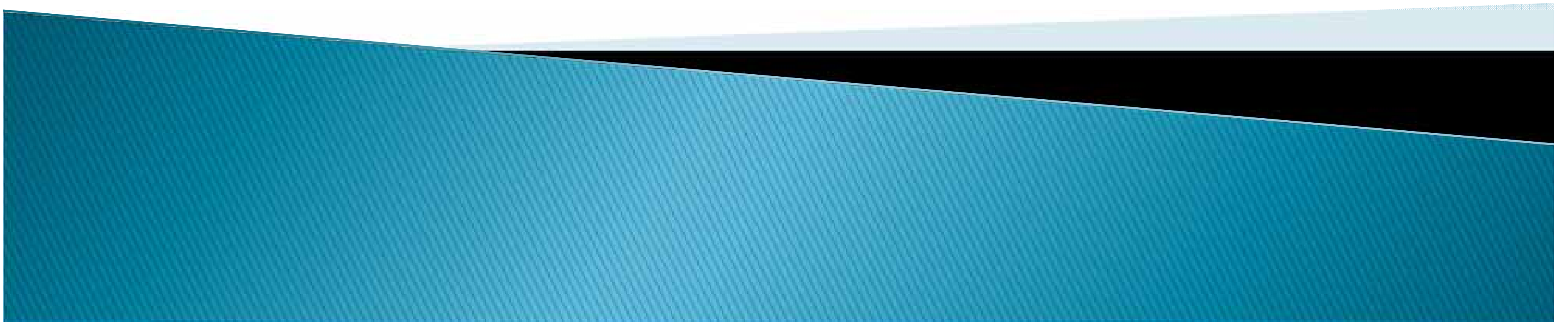
- Normal VA; Normal pupil reaction
- Lens/Cornea clear
- Extra-ocular movements: restricted lateral gaze BL
- Diplopia in primary position and temporal gaze
- Fundus: BL disc oedema
- Conclusion: Papilloedema & BL LR palsy

Management – 2

- Diagnosis:
  - Neck stiffness, headache, upper arm pain, LR palsy, papilloedema
  - Normal brain imaging; ? CSF
  - Obese (wt=60kg)
  - *Benign Intracranial Hypertension*
- Dexamethasone 4mg q6H
- Diamox 500mg BD
- Remarkable improvement after 24 hrs: all symptoms resolved. Diplopia improving.
- Discharged home 23.08.13 on solupred & diamox
- OPD review: Diplopia on far vision only

Benign Intracranial Hypertension

A Paediatric Perspective



Introduction

Definition

- Increased intracranial pressure without a SOL or hydrocephalus and normal CSF composition
- Pseudotumour cerebri
- Idiopathic intracranial hypertension (IIH)

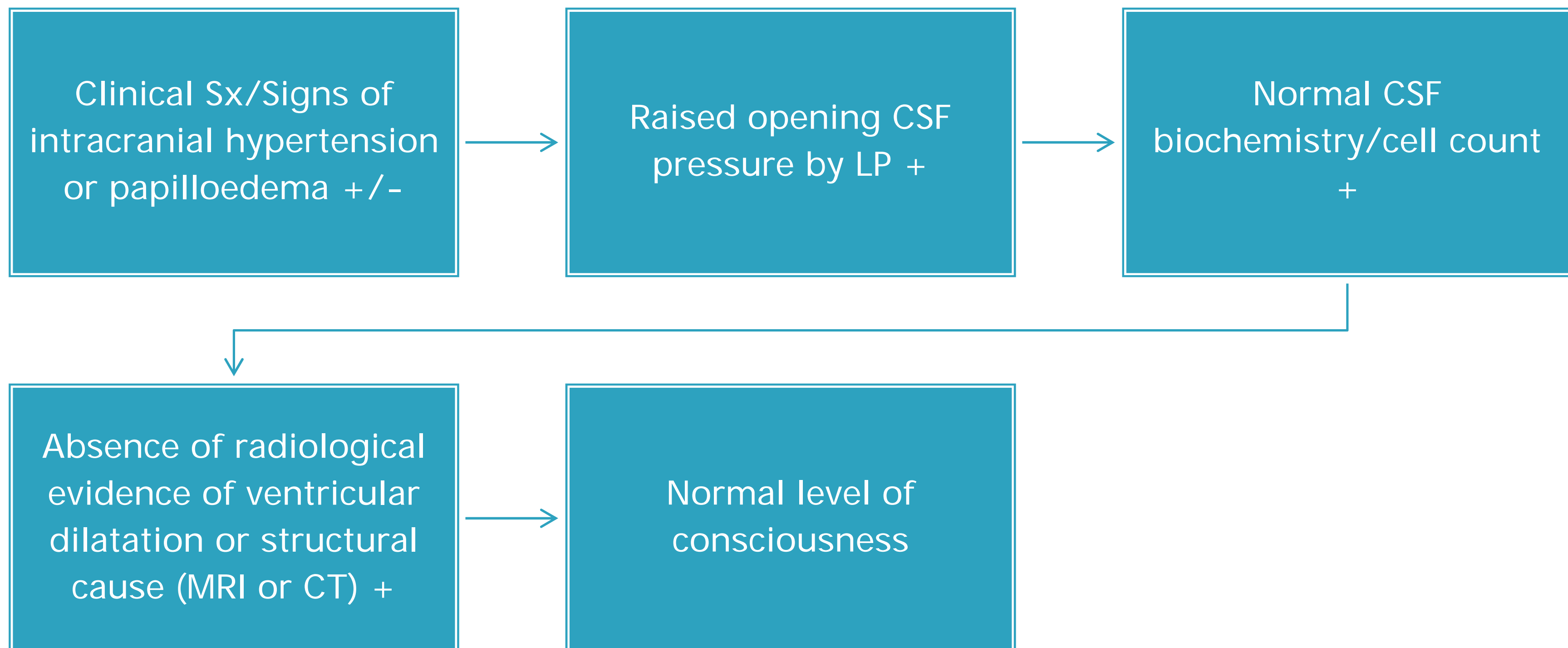
Incidence

- Overall annual incidence: 1-3/100,000
- Children: 0.1-0.9/100,000

Aetiology

- Adults and post-pubertal children: obesity; female gender
- Pre-pubertal children: no association with gender or obesity

Diagnostic criteria



Symptoms/Signs of Intracranial hypertension

↑ICP

- Headache
- Nausea/vomiting
- Irritability, lethargy, dizziness
- Neck stiffness
- Facial paresis, back/neck pain, radicular pain
- Tinnitus

Visual Sx

- Papilloedema (UL or BL)
- Transient visual loss
- Diplopia
- Squint; 6th nerve plasy
- Visual acuity loss
- Visual field defect

Associations

Drugs

- Penicillins
- OCPs
- Steroids
- Isotretinoin

Endocrine/Metabolic

- Obesity
- Hypo/hyper thyroidism
- Cushing's
- POCS
- Menarche

Systemic disorders

- SLE
- Sickle cell anaemia
- Migraine
- Lyme disease
- HIV

Management

Rectifying underlying predisposing condition/ withdrawal precipitating factor

Therapeutic LP

Medical Rx: Analgesics, Carbonic anhydrase inhibitors, Diuretics , Corticosteroids

Surgery: CSF diversions; Optic nerve sheath decompression

Medical Therapy

Acetazolamide

- Inhibits CSF production/secretion
- 25mg/k/d →100mg/k/d
- Wean after 2 months

Frusemide

- 1-2 mg/kg/D
- Loop diuretic

Steroids

- Side effects
- Wean after 2 weeks
- PDN:1-2mg/k/d
- Dexamethasone 0.1-0.75 mg/k/d

Prognosis

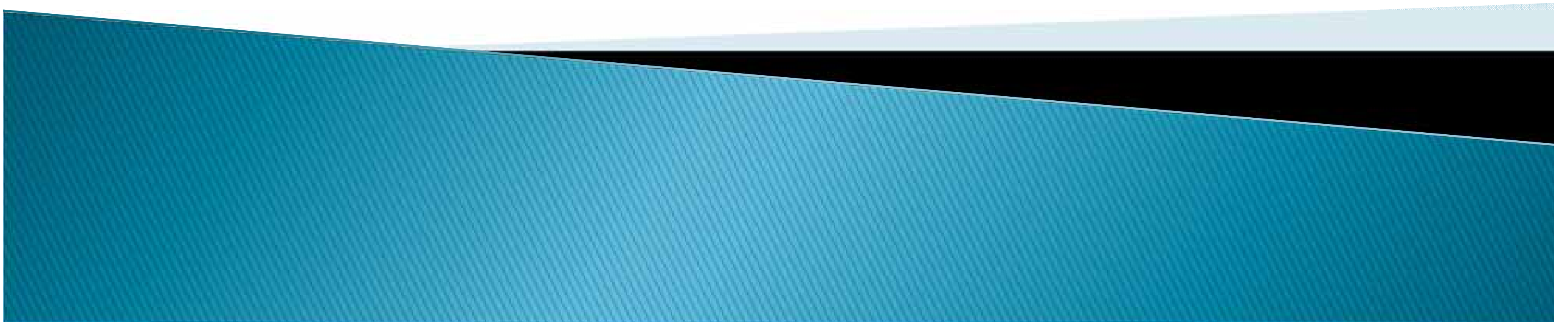
Initial remission: 1w-6m

Recurrence rates: 40%

Visual outcome: spontaneous resolution v/s long term visual impairment

Case Presentation – 2

A pain in the foot……



History

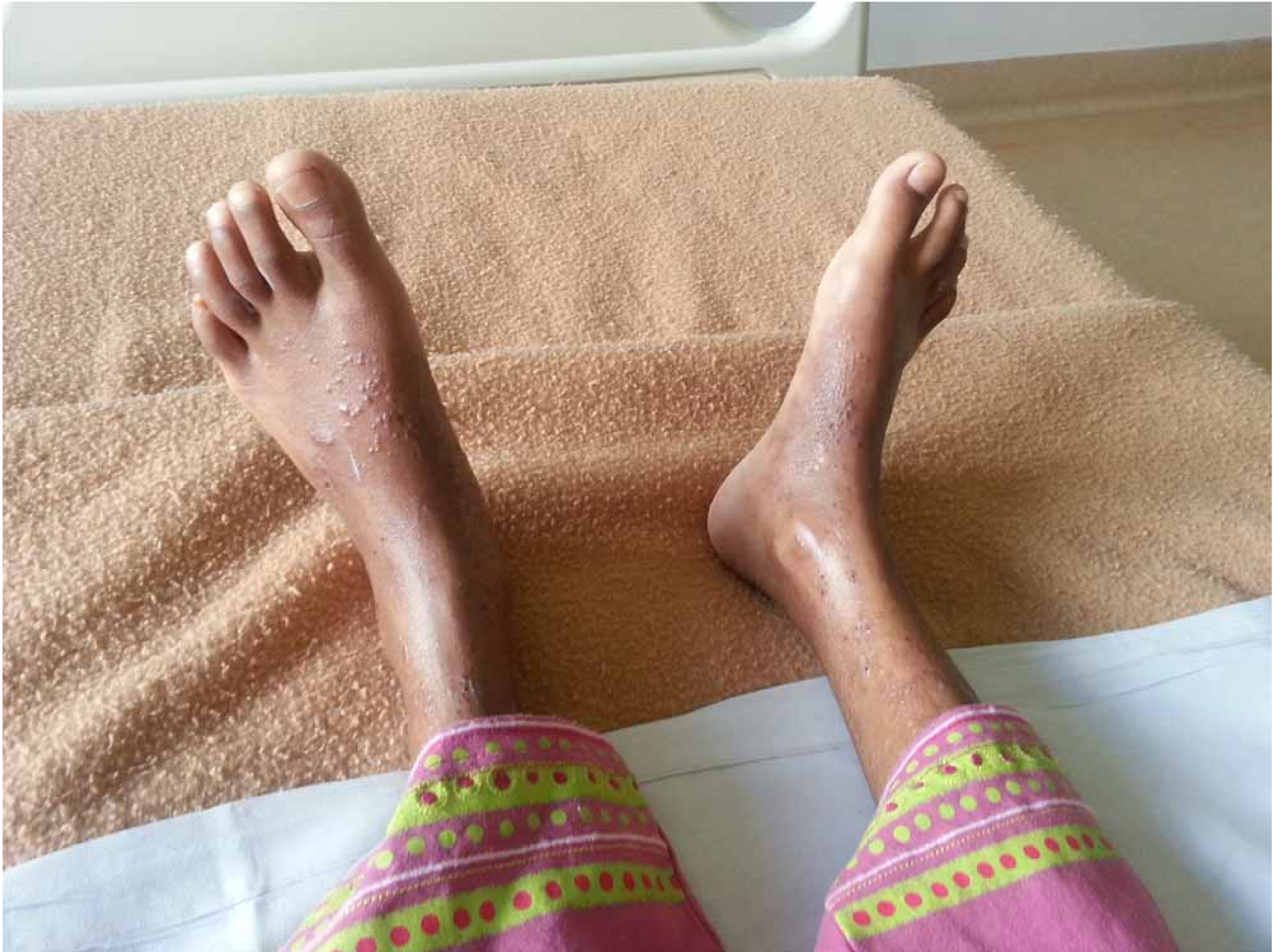
- Y.A – Female - 9 years
- May 2013: Fall at school. Pain ankles L>R + intermittent fever
- Orthopaedic opinion 2 weeks after onset: x-ray – no fracture – ibuprofen syrup (4/7)
- Admitted SSRNH for 24 hrs – CRP/ESR ↑
- ?JRA – PDN started (2/7)
- RF & Anti-CCP negative

History

- OPD ABH 30.07.13
- May 2013: rash on forearms/feet/cheeks – vesicular; Lip swelling/Bleeding mouth ulcers
- July 2013: peri-anal abscesses; severe constipation; bleeding PR

Examination

- Wt 20.6kg
- Massive faecal loading
- Vesicular/Pustular rash on feet with punched-out lesions.
- Healed lesions cheeks & forearms with scars
- Swollen/tender lt ankle
- Multiple peri-anal large pustules
- Healing ulcers over labia majora



Investigations

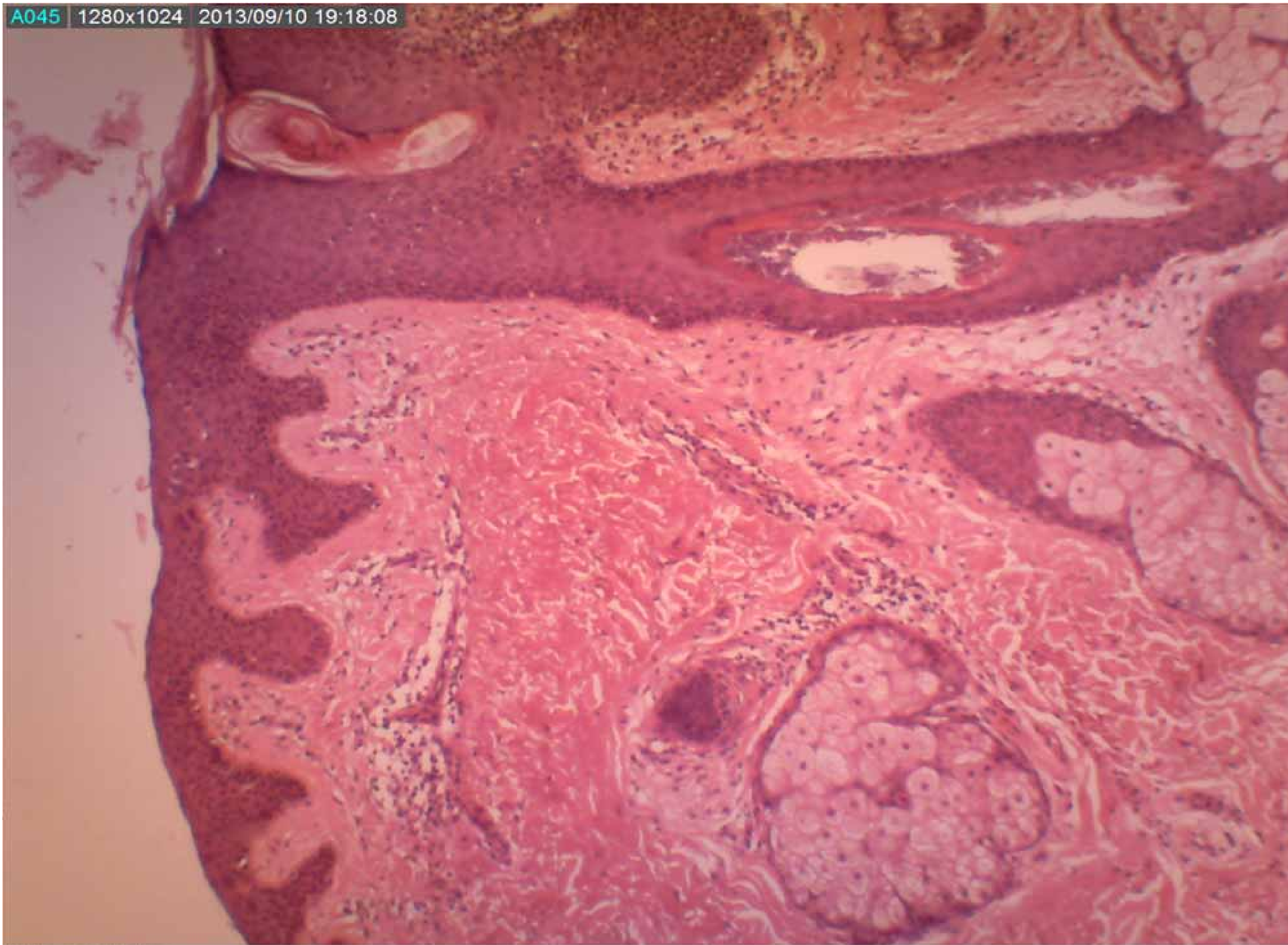
- MRI ankle joint: small amount of intra-articular fluid
- AXR: Massive faecal loading
- Hb 10.2. Iron & Transferrin: N. Ferritin ↑
- TLC 12.84 (N 73%); Platelets 644; ESR 115
- ASOT <200; Anti-CCP –ve; LFTs N; CRP 62.6

Differential Diagnosis

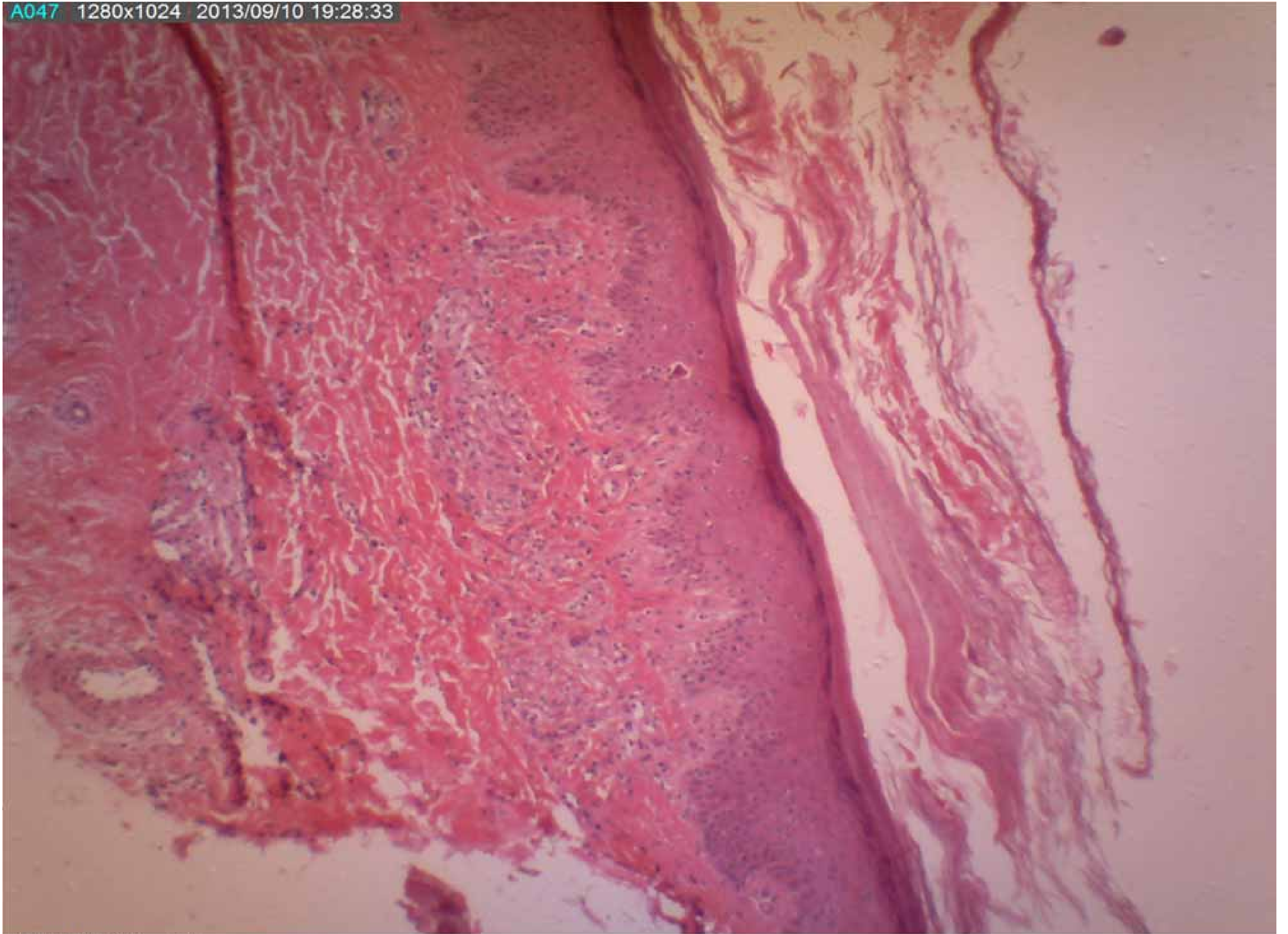
	Crohn's Disease	Behcet's Disease
Arthritis	+	+
Mouth ulcers	+	+
Genital ulcers	-	+
Peri-anal lesions	abscesses	ulcers
Rash	Erythema nodosum Pyoderma gangrenosum	Erythema nodosum Pseudofolliculitis Papulo-pustular lesions
Constipation	+/-	+/-
Bleeding PR	_	+

Management

- Macrogols
- Topical anti-septic & oral azithromycin
- Manual evacuation under GA
- Skin biopsy: Peri-anal and ankle



Skin-no pathology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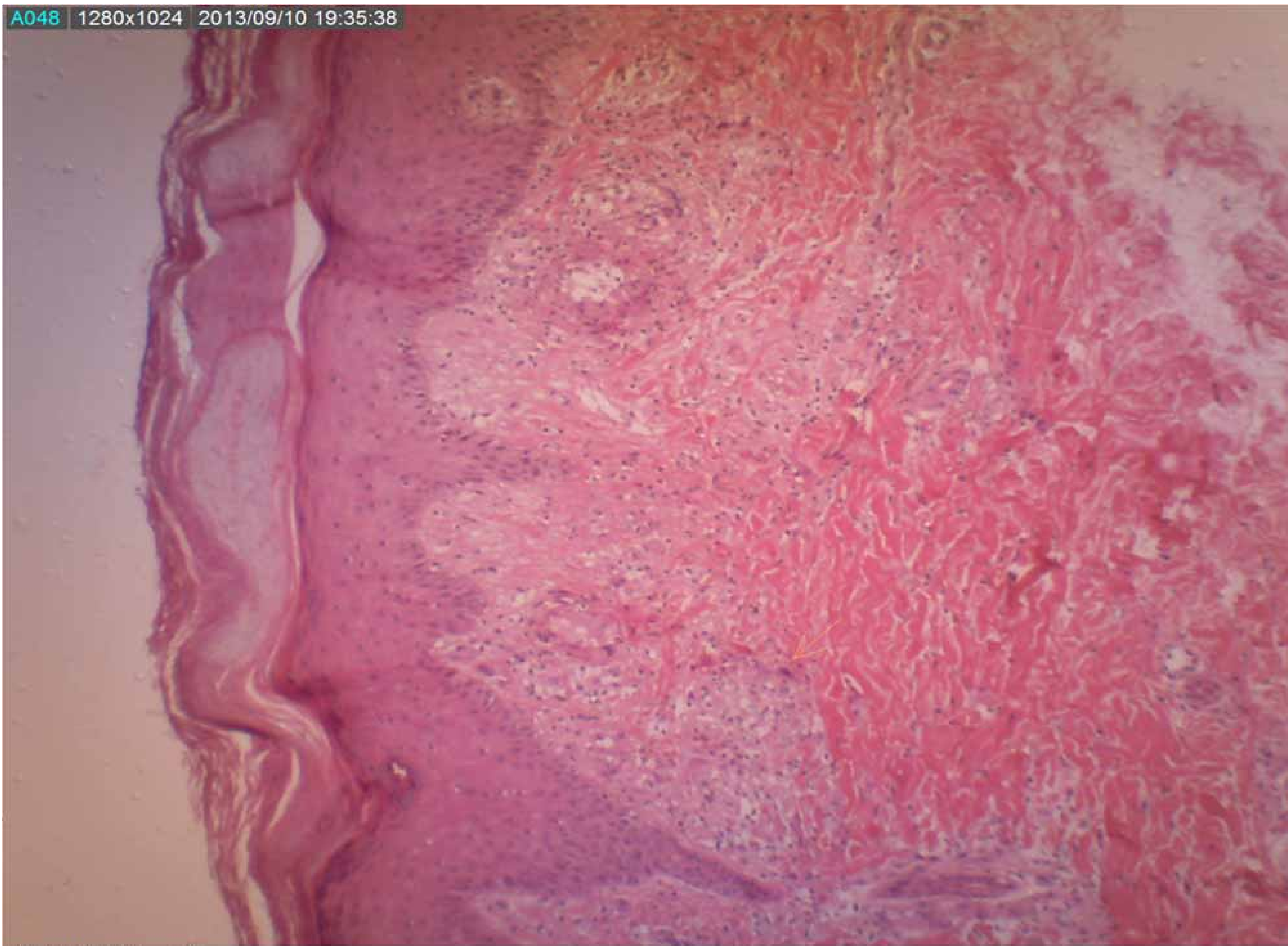
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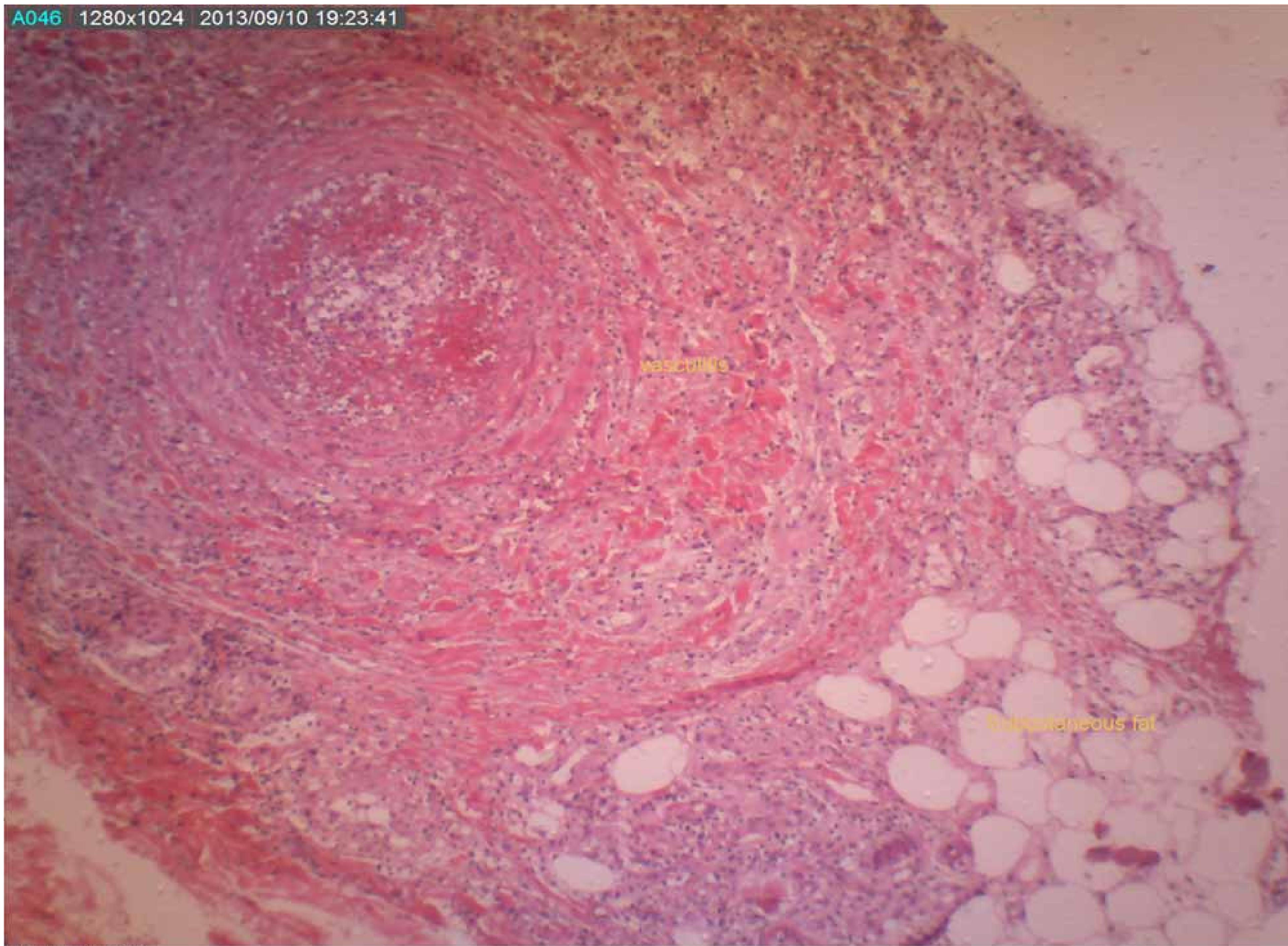

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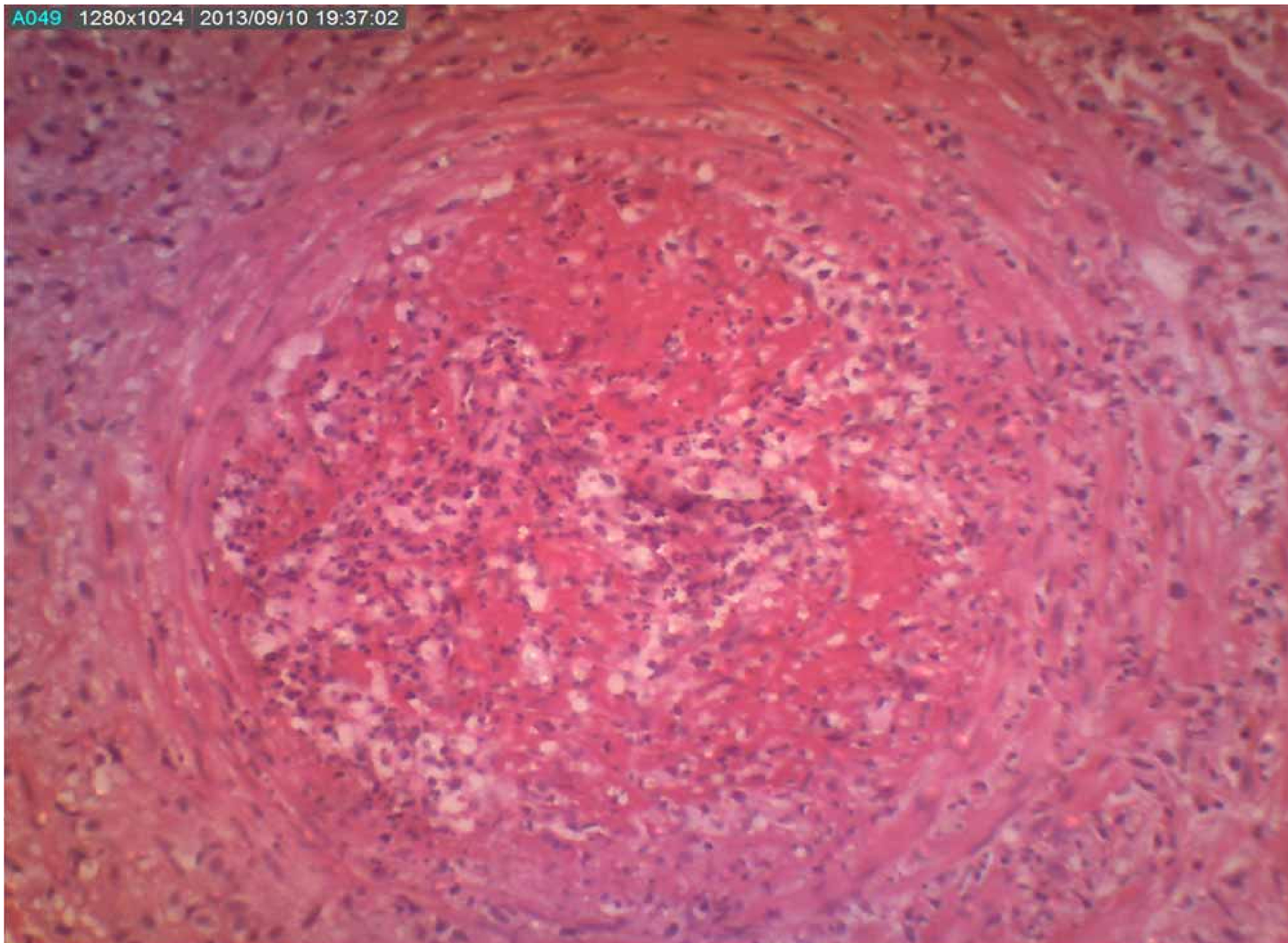
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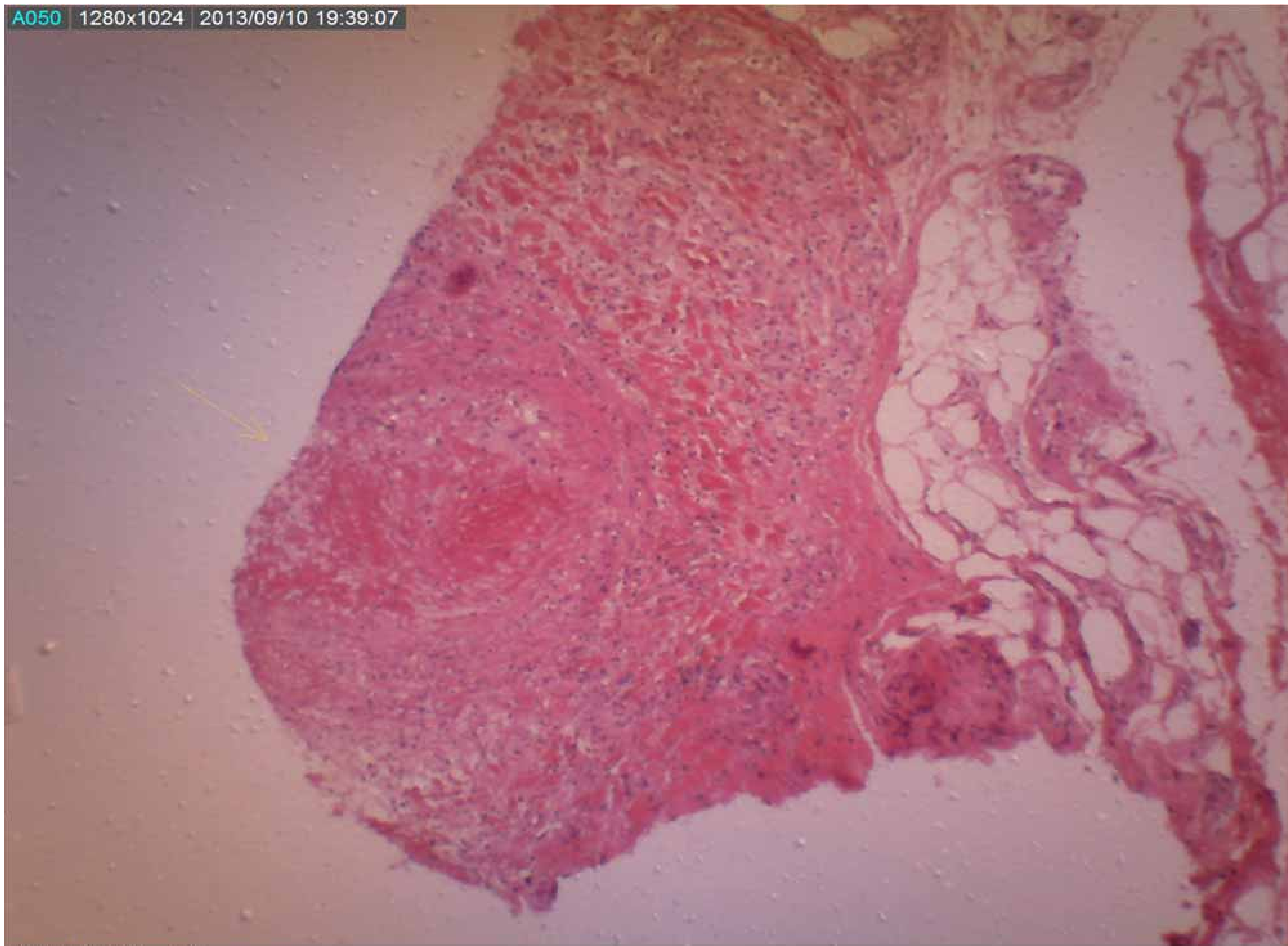

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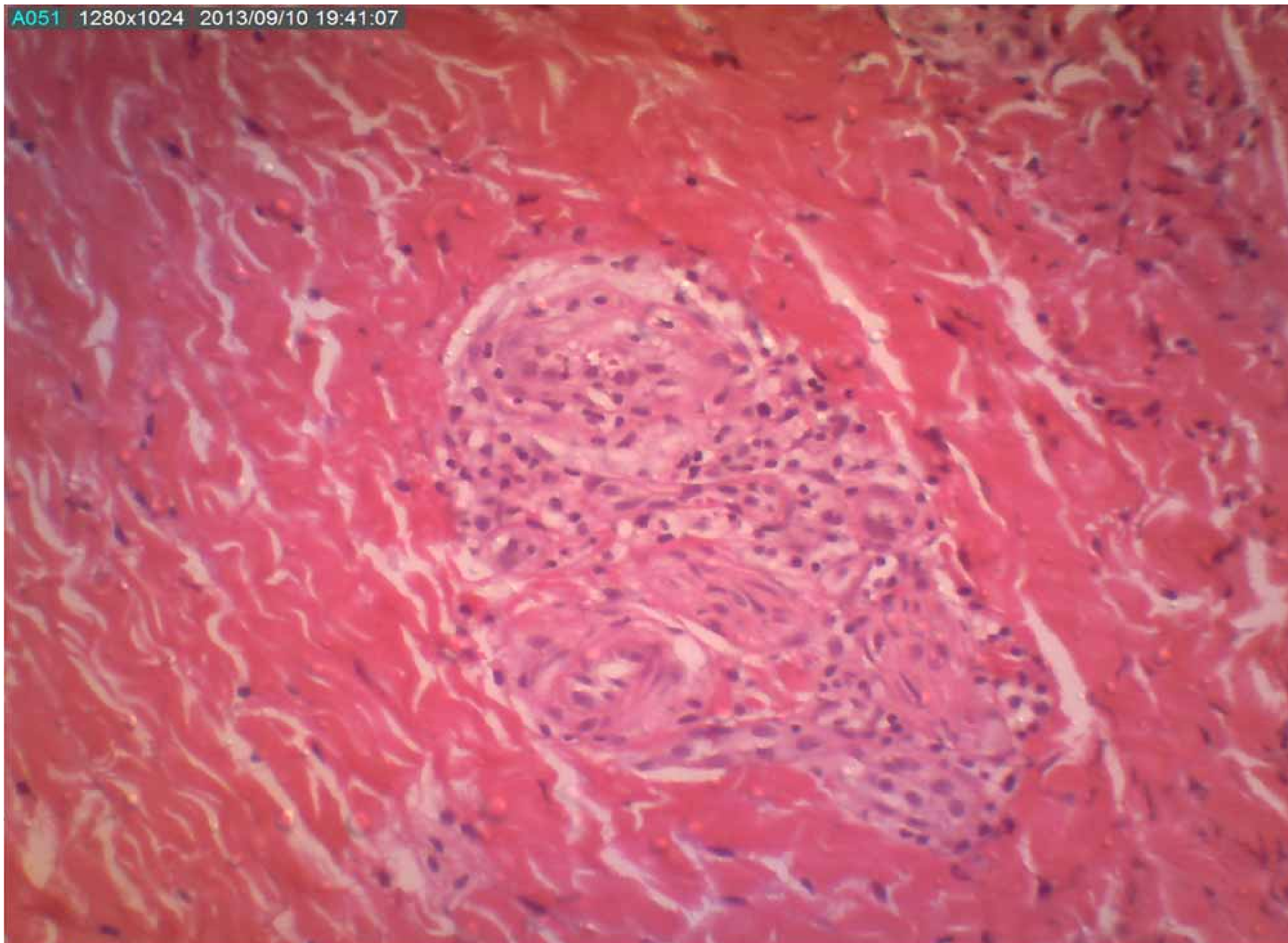

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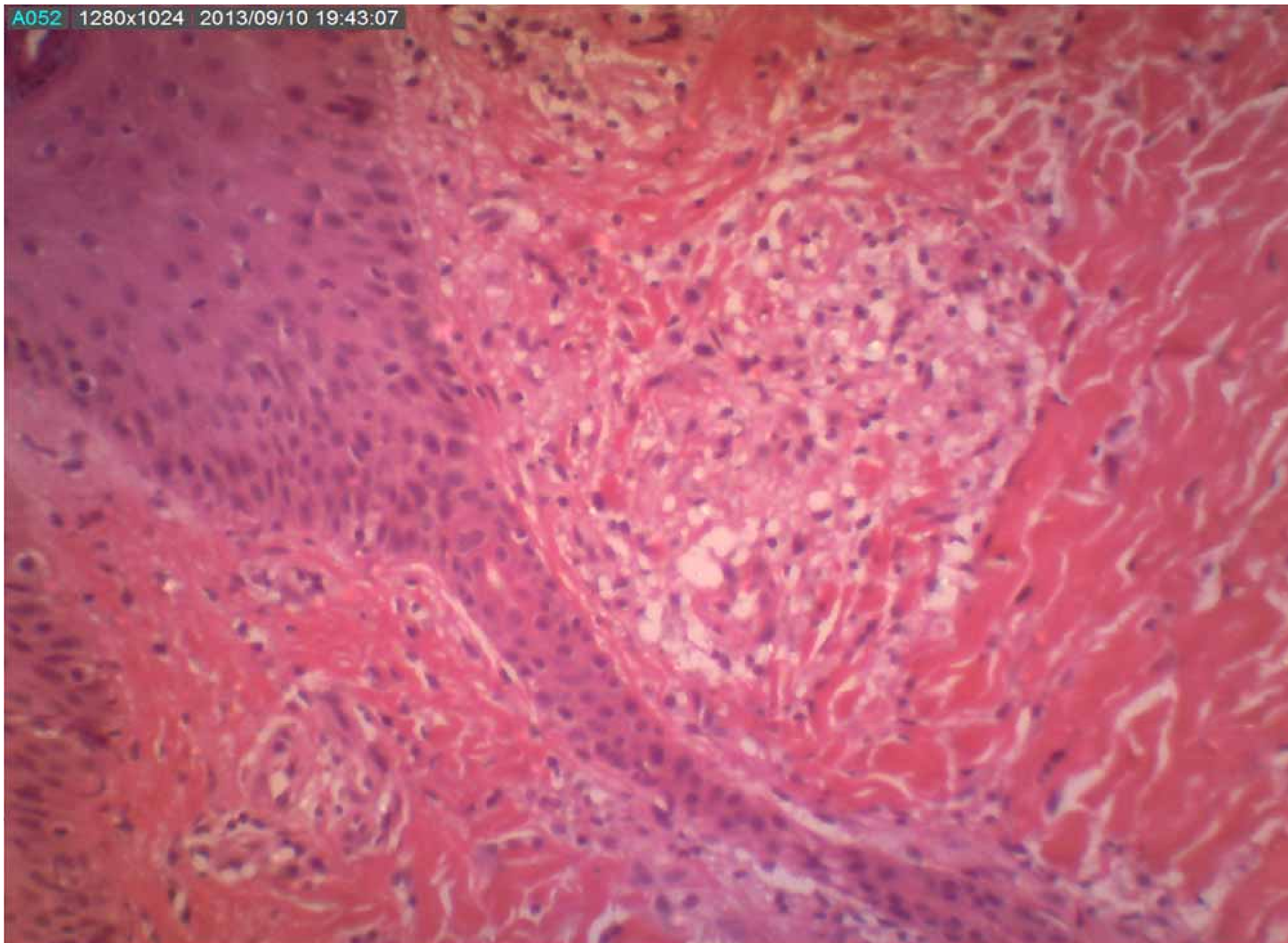
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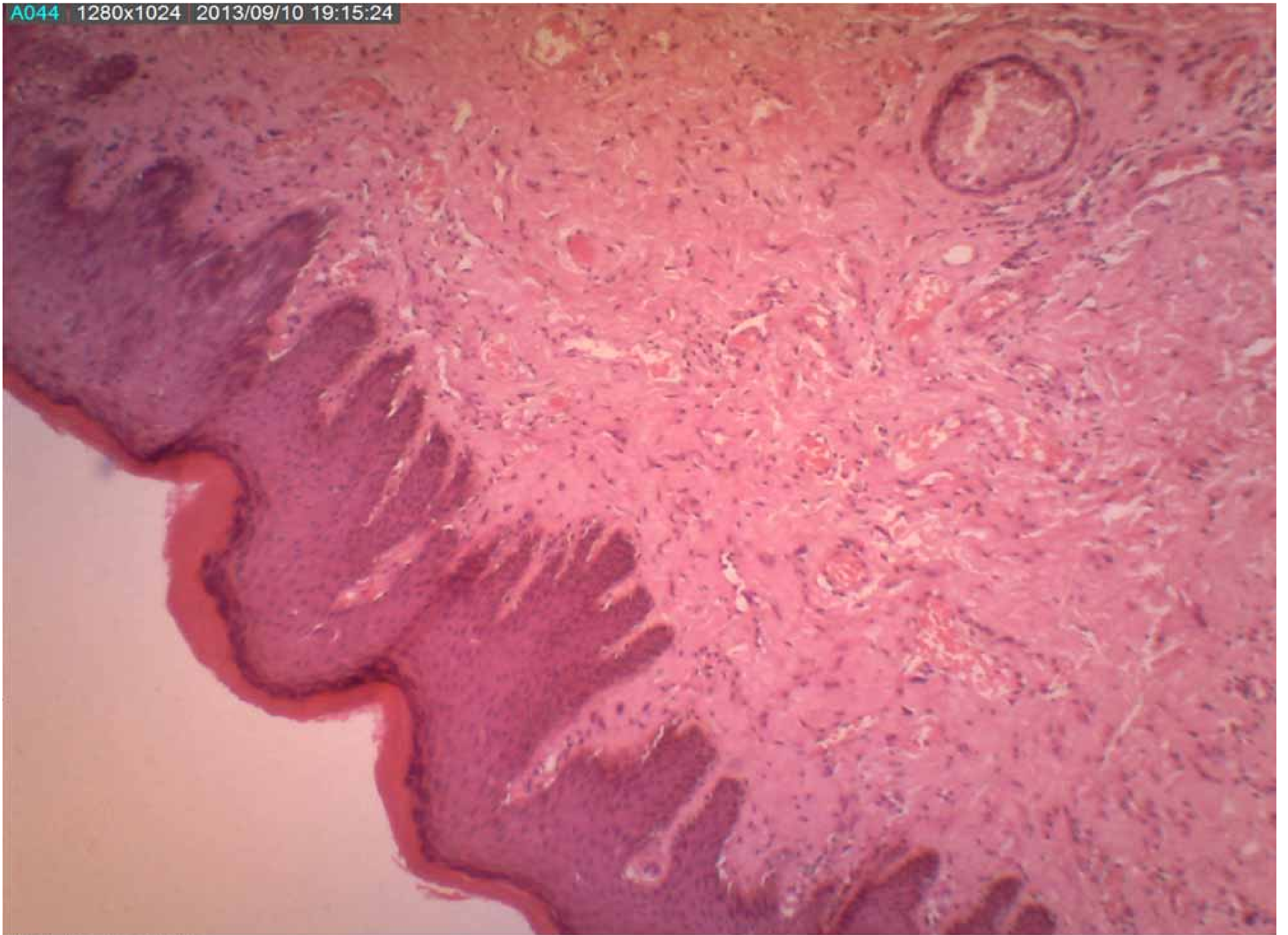
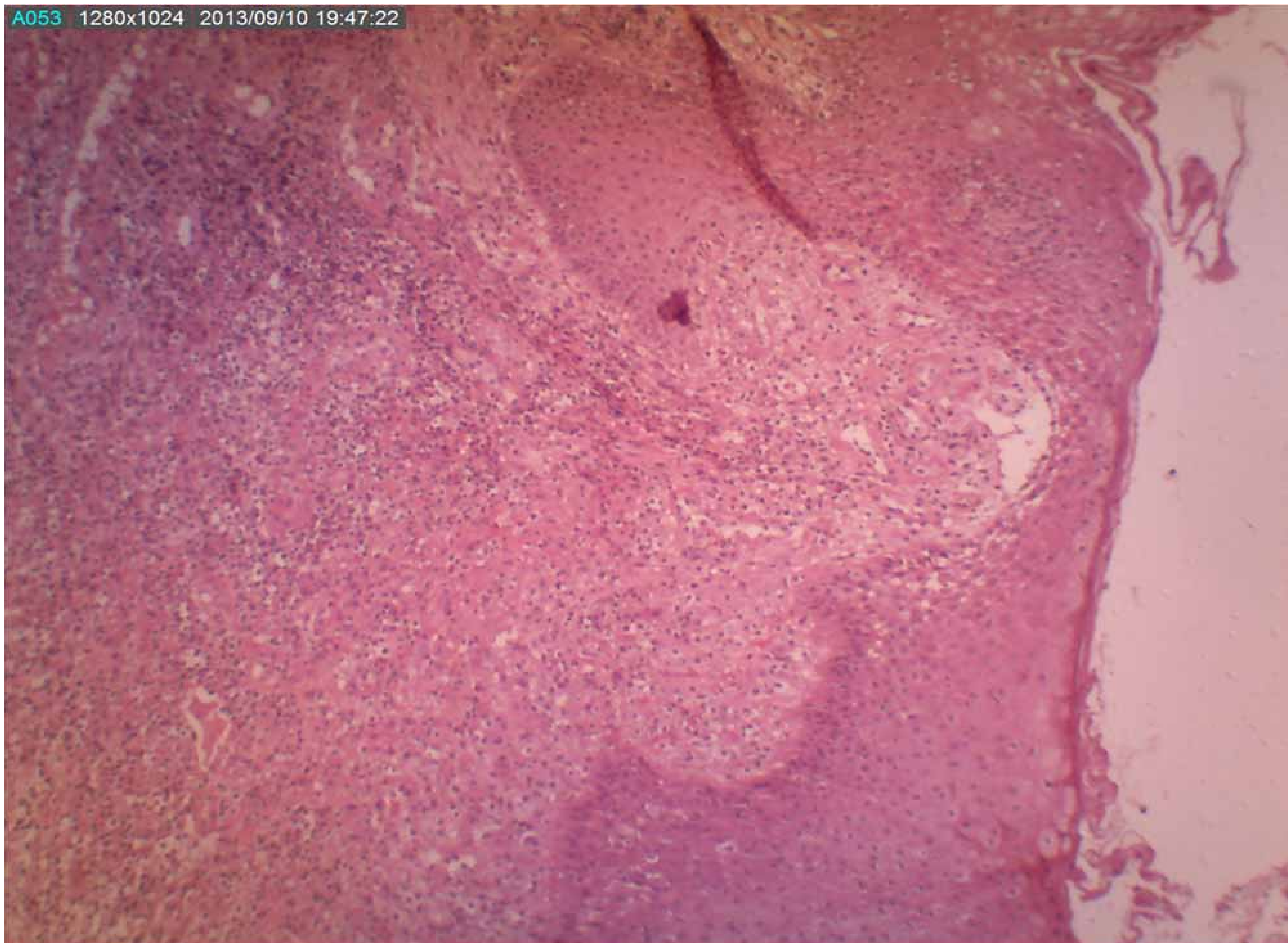


Anal skin-no pathology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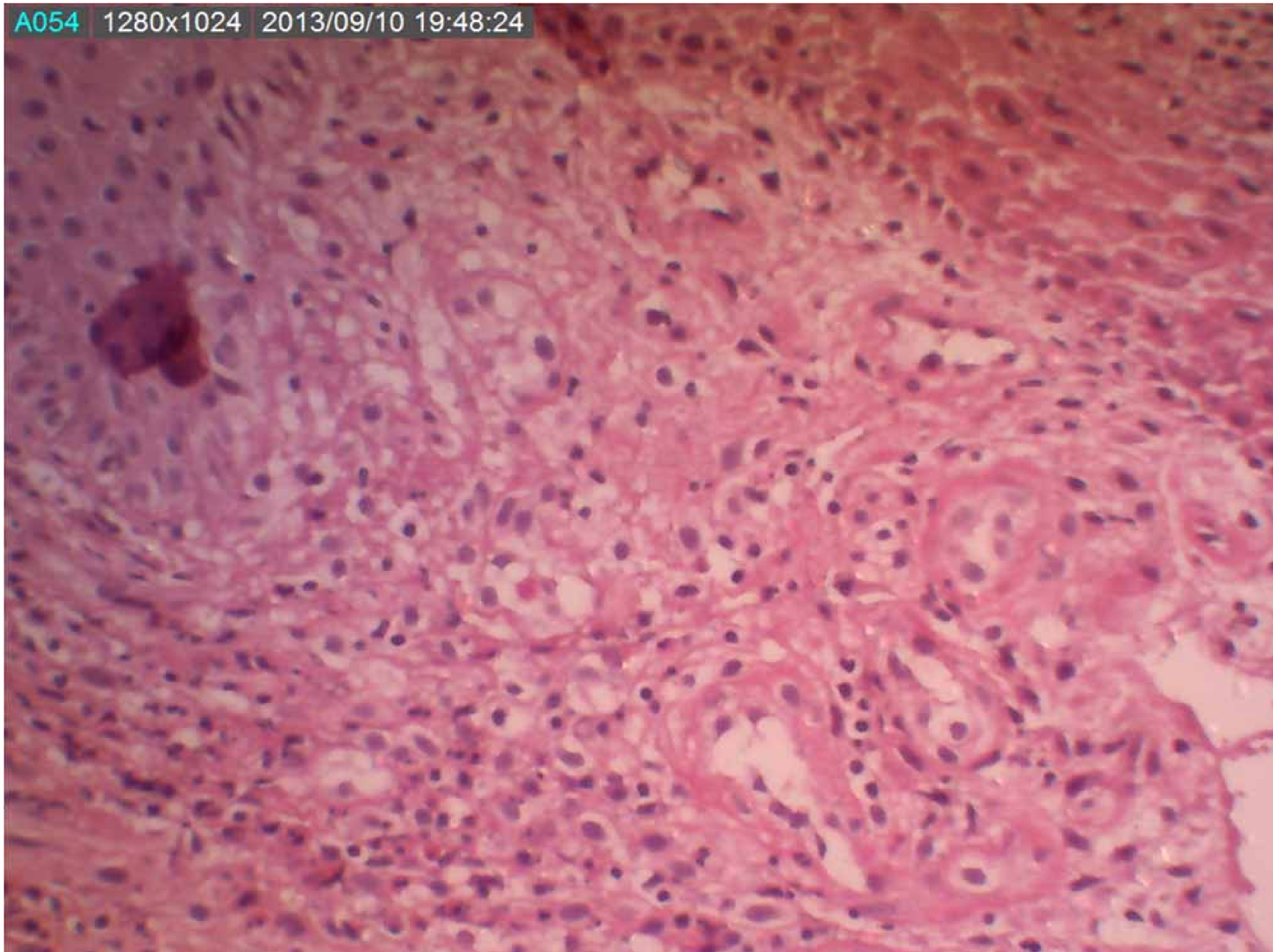
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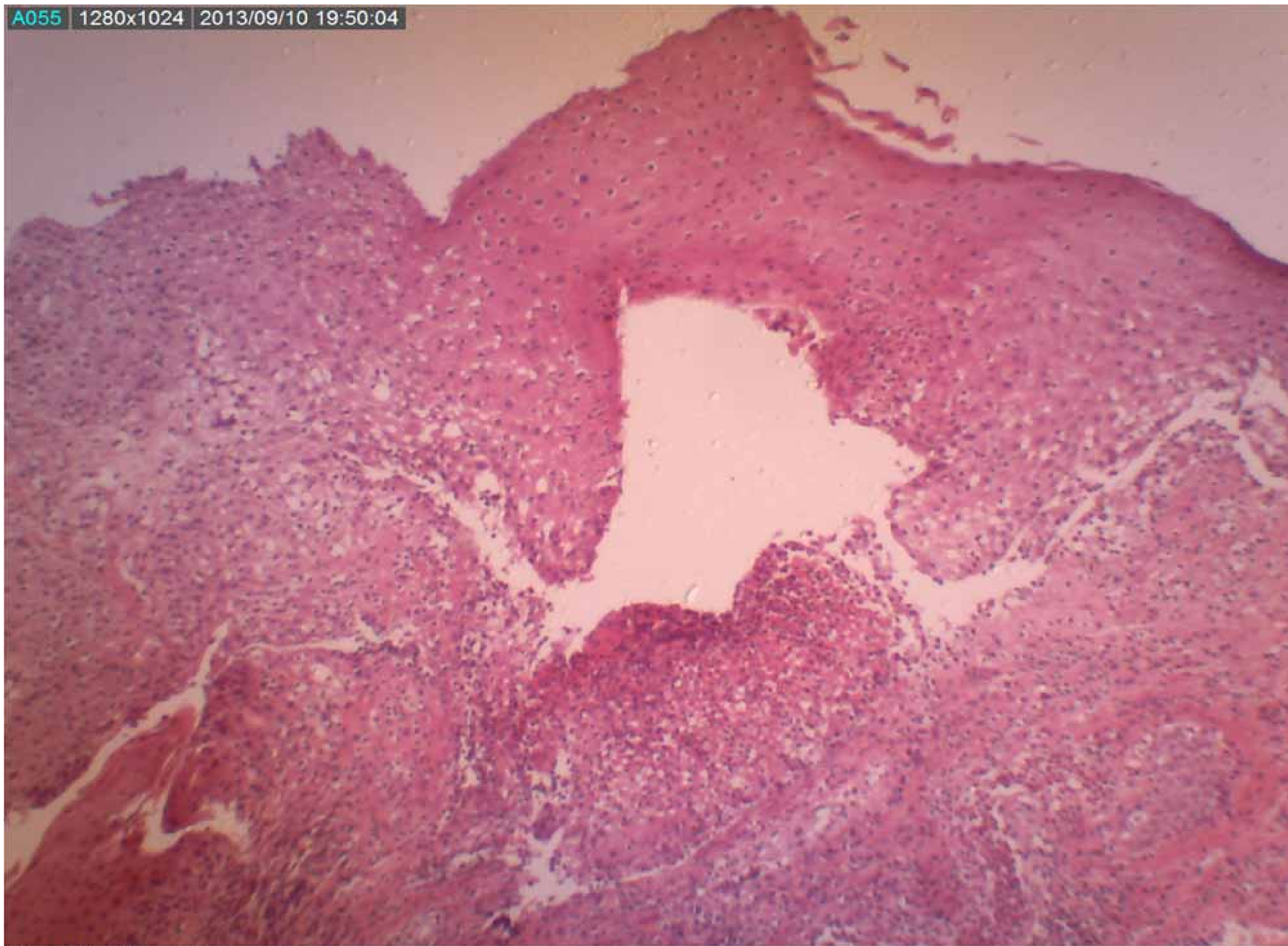
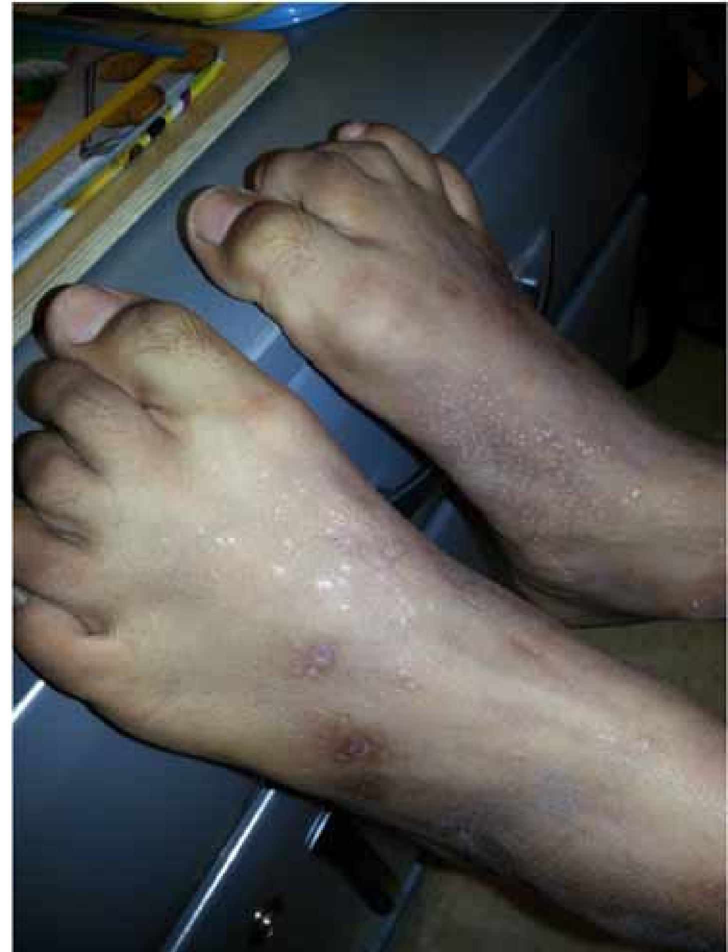


ABH-H13/684B x40

Management

- Diagnosis: ?Behcet's Disease
- Oral PDN started
- Discharged 05.08.13
- OPD review
- ESR 27 CRP 1.1



Behcet's Disease in children

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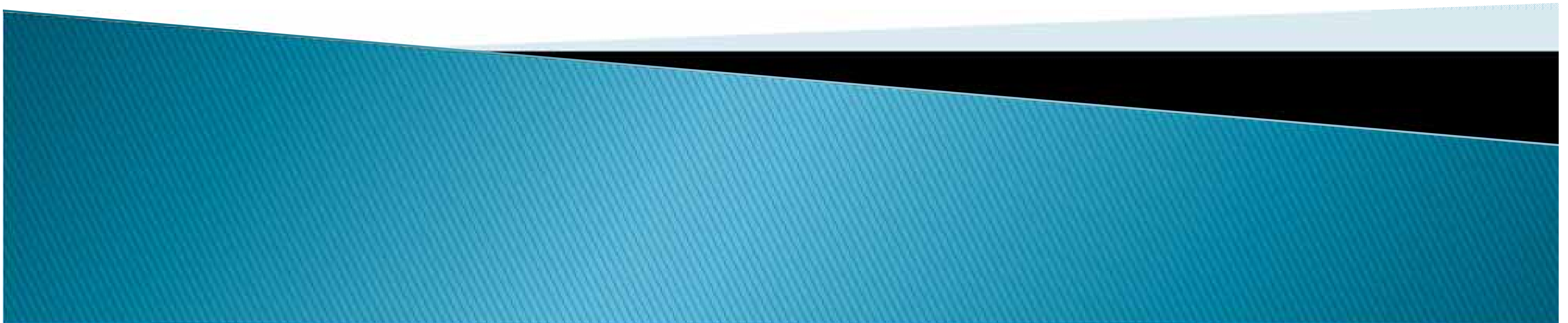

Introduction

Systemic vasculitis with multi-organ involvement

Highest incidence along the Old Silk Road: Turkey, Middle East, China, Japan

Children: Age of onset 4 yrs; Age at diagnosis 11 yrs

Diagnostic Criteria

- 3 episodes of oral herpetiform or aphthous ulcerations within a 12-month period + 2 of the following:
- Recurrent painful genital ulcers that heal with scarring
- Ophthalmic lesions, including anterior or posterior uveitis, hypopyon, or retinal vasculitis
- Skin lesions, including erythema nodosum–like lesions, pseudofolliculitis, or papulopustular or acneiform lesions
- Positive results from pathergy skin testing, defined as the formation of a sterile erythematous papule 2 mm in diameter or larger that appears 48 hours following a skin prick with a sharp sterile needle (22-24 gauge [a dull needle may be used as a control])

Other manifestations

- Joints: Arthritis and arthralgias occur in as many as 60% of patients and primarily affect the lower extremities, especially the knee. Ankles, wrists, and elbows can also be primarily involved.
- GIT: Symptoms include abdominal pain, bloating, peri-anal ulcers and GI bleeding.
- CNS/Vasculopathy

Treatment

Depends on

- Severity of disease
- Organ involvement

Drugs

- NSAID/Systemic steroids
- Immunosuppressants
- Sulfasalazine
- Thalidomide!

THANK YOU

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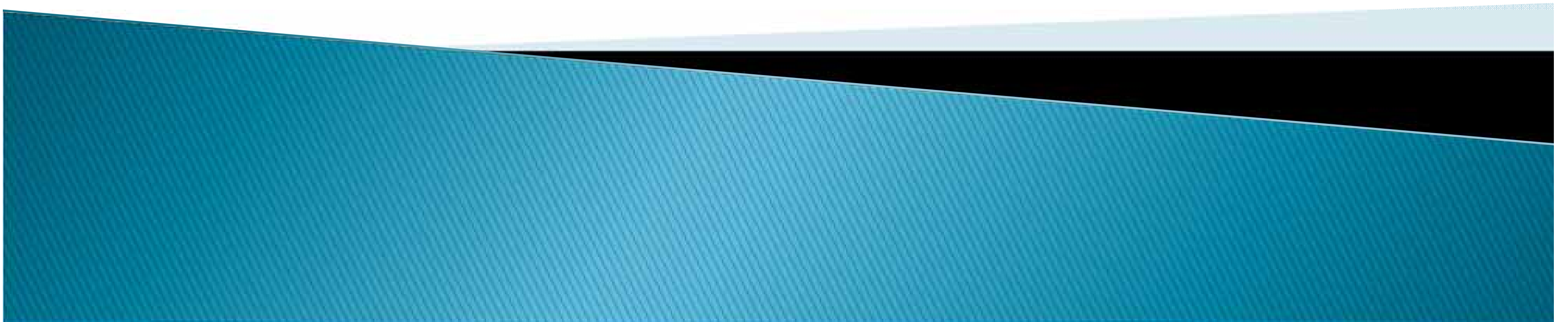
"What do you mean you're the new Paediatric Specialist?"

Paediatric Crohn’s Disease



Clinical features

Only 25% present with classical triad of abdo pain, wt loss, diarrhoea.

Extra-intestinal manifestations dominate clinical presentation.

Lethargy, anorexia, fever, nausea, vomiting, arthropathty, malnutrition

Erythema nodosum, pyoderma gangrenosum, arthritis, oral disease (lip swelling, aphthous ulcers), peri-anal disease (skin tags, fistulae, abscesses)

Investigations

Blood tests

- FBC, LFTs, ESR, CRP
- ASCA, pANCA

Endoscopy

- Upper GI
- Total colonoscopy
- + Biopsy

Imaging

- MRI enteroclysis
- Capsule endoscopy
- MRI of pelvis

Management

First line induction of remission	• Enteral nutrition • Corticosteroids
2ND line maintenance	• Azathioprine • 6-MP
3RD line therapy	• Anti-tumour necrosis factor (Infliximab) • ?surgery